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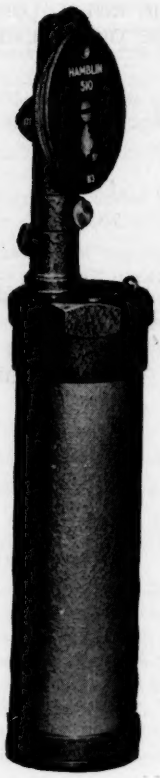

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Brit. J. Ophthal. (1958) **42**, 385.

COMMUNICATIONS

THE EFFECT OF CERVICAL SYMPATHOTOMY ON AN OCULAR RESPONSE TO HYPOTHALAMIC STIMULATION*

BY

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RECENT work (von Sallmann and Lowenstein, 1955; Gloster and Greaves, 1956, 1957) has shown that in the cat electrical stimulation of the diencephalon is accompanied by changes in intra-ocular pressure. Many of these effects can be attributed to simultaneous changes in the systemic blood pressure, but in a previous study we demonstrated that a reduction in intra-ocular pressure could occur with a *rise* in blood pressure and was accompanied by bilateral pupillary dilatation with occasional retraction of the nictitating membrane. This type of response was obtained from a limited area in the medial hypothalamus near the anterior column of the fornix (Gloster and Greaves, 1956, 1957). We suggested that this response of the intra-ocular pressure was due to a reduction of capillary blood pressure within the eye. However, the accompanying rise in systemic blood pressure, which probably results from a general sympathetic discharge, would ordinarily cause a rise in capillary blood pressure, and it is therefore necessary to postulate that this tendency is outweighed by a constriction of the ocular blood supply, which is possibly due to local sympathetic activity. This hypothesis can be tested by examining the effect of cervical sympathotomy on the response.

It is the purpose of this paper to describe the response in greater detail and to show that the results of cervical sympathotomy do, in fact, support our hypothesis.

Methods

The experimental procedure followed in this study was essentially the same as that described fully in a previous paper (Gloster and Greaves, 1957). Cats were anaesthetized by intravenous injections of chloralose (100 mg./kg.), the femoral blood pressure, intra-ocular pressures, and nictitating membrane responses being recorded continuously on a photographic film (Greaves and Perkins, 1952). In five animals a portion of the preganglionic cervical sympathetic nerve was exposed on each side of the neck and carefully separated from the vagus for about 5 mm., a thin silk thread being passed round the nerve to allow identification when required. After the head of the animal had been mounted in the stereotaxic apparatus, the stimulating electrode was placed in a suitable position in the medial part of the hypothalamus and moved downwards in steps of 1 mm. until the

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characteristic response was obtained on stimulation. After the response had been recorded, preganglionic cervical sympathotomy was performed on the side of stimulation. Stimulation was then repeated without changing the position of the electrode. In three animals the cervical sympathetic nerve was then cut on the side opposite to that of stimulation and the response to a further stimulation was recorded. In one animal the blood vessels of the ear on the side of stimulation were observed through a microscope before, during, and after the application of each stimulus.

At the conclusion of the experiment the brain was removed and sectioned in order to determine the position of the tip of the electrode.

Results

(A) Characteristics of the Response

In its most easily recognizable form the response consists of:

(1) A fall in intra-ocular pressure in the ipsilateral* eye.
(2) A rise in systemic blood pressure.
(3) Retraction of the nictitating membrane.
(4) Pupillary dilatation.

The constant feature of the response is the fall in intra-ocular pressure occurring in the ipsilateral eye, the other components showing some variation. The following account is derived from experiments on fourteen animals.

The fall in intra-ocular pressure in the ipsilateral eye generally amounted to 1 to 3 cm. saline, although a fall of 6 cm. was noted on one occasion. Fig. 1*a* (opposite) is a tracing showing the main features of the response. Sometimes a small temporary elevation of intra-ocular pressure was found at the beginning of stimulation, with a similar depression at the end (see Fig. 4, below). The fall in intra-ocular pressure usually lasted throughout the period of stimulation but was occasionally transient. In ten of the fourteen animals the intra-ocular pressure also fell in the contralateral eye but not usually to the same extent as in the ipsilateral eye.

A rise of 5 to 65 mm. Hg in blood pressure accompanied the fall in intra-ocular pressure in eleven out of fourteen animals. In one animal the blood pressure fell (Fig. 1*b* opposite), and the other two showed complex responses in which a slight fall in blood pressure was preceded by a short but definite rise.

Retractions of the ipsilateral nictitating membrane occurred in eight animals, and in three of these the effect was bilateral, although the response of the contralateral membrane was weaker.

Pupillary dilatation occurred in all but one animal. In eight animals the dilatation was equal on the two sides, but it was greater in the ipsilateral eye in the remaining five. The dilatation was brisk, commencing usually within

* The word "ipsilateral" is used throughout this paper as denoting the same side as that of hypothalamic stimulation; "contralateral" always refers to the side opposite to that of stimulation.

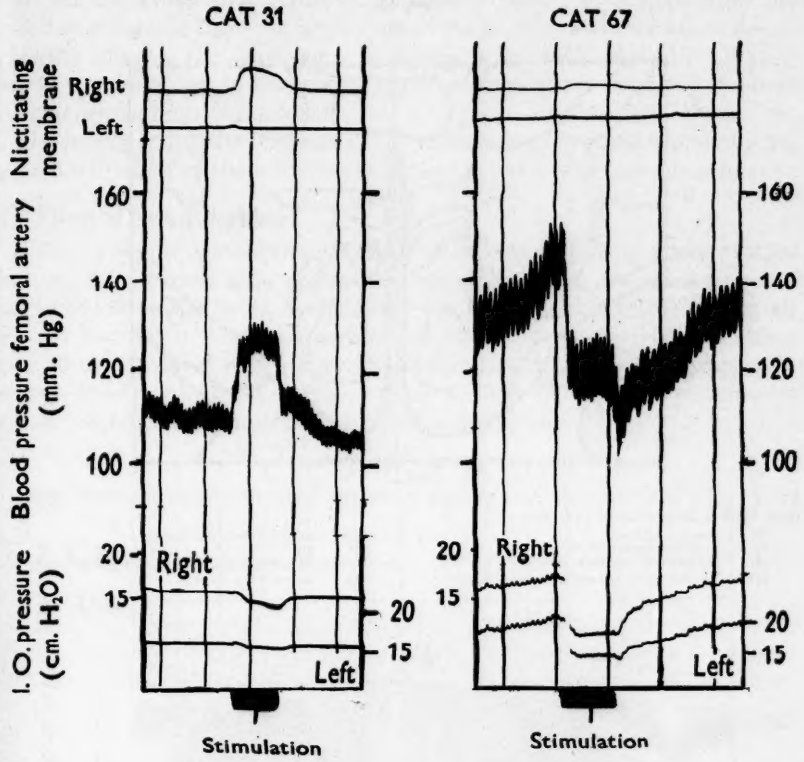

FIG. 1 (*a*).—Tracing showing fall in intra-ocular pressure with a simultaneous *rise* in blood pressure. Note also retraction of right nictitating membrane.

FIG. 1 (*b*).—Tracing showing large bilateral fall in intra-ocular pressure with accompanying fall in blood pressure. Note also retraction of right nictitating membrane.

Interval between vertical lines = 60 sec. Localization: A 10, R 1·5, H + 5½.

1 second of the onset of stimulation, reaching a maximum in about 5 to 10 seconds, and lessening after 40 to 45 seconds. The amount of dilatation was usually 6 to 8 mm., reaching 9 to 10 mm. in three animals.

(B) Localization of Effective Stimuli

This response has been obtained from the stimulation of points situated mainly in the medial hypothalamus in frontal planes 10 to 13 mm. anterior to the inter-aural line (A10 to A13). It appeared that the response was most frequently obtained from points in A12 and their distribution is shown in Fig. 2 (overleaf); a section of the brain in one of these experiments is *illustrated in Fig. 3 (overleaf).*

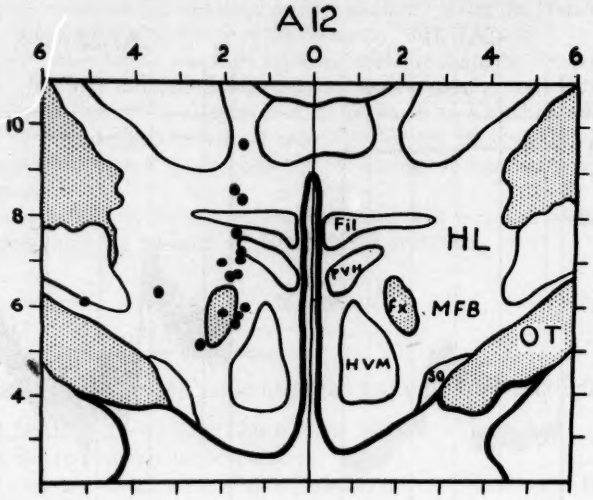

FIG. 2.—Map of frontal plane A 12 showing points from which stimulation produced independent falls in intra-ocular pressure.

Fil	Nucleus filiformis	MFB	Medial forebrain bundle
Fx	Anterior column of fornix	OT	Optic tract
HL	Lateral hypothalamic area	PVH	Paraventricular nucleus of hypothalamus
HVM	Ventromedial nucleus of hypothalamus	SO	Supra-optic nucleus

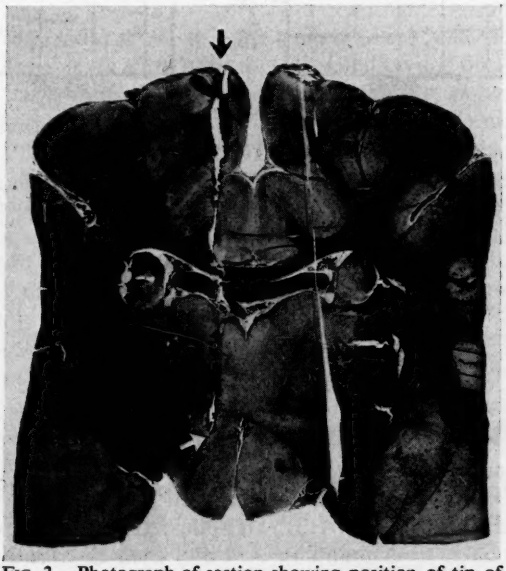

FIG. 3.—Photograph of section showing position of tip of electrode in one of the experiments indicated in Fig. 2. Arrows show extent of electrode track.

It will be noted that most of the points lie 1·5 to 2 mm. from the mid-line extending from the lower part of the anterior column of the fornix upwards towards the thalamus. In planes A10, A11, and A13, all such points were confined to the medial hypothalamus in the neighbourhood of the anterior column of the fornix.

The points giving the response in the contralateral eye had essentially the same distribution as those for the ipsilateral eye but were less numerous.

(C) Effect of Sympathotomy

Changes in the characteristics of the response following acute sympathotomy on the same side as hypothalamic stimulation are summarized in the Table; one experiment is reproduced in Fig. 4. This shows that in all animals the fall in intra-ocular pressure in the ipsilateral eye was either reduced or converted into a rise after sympathotomy. No such consistent change was found in the contralateral eye. The blood pressure response was not significantly affected by sympathotomy.

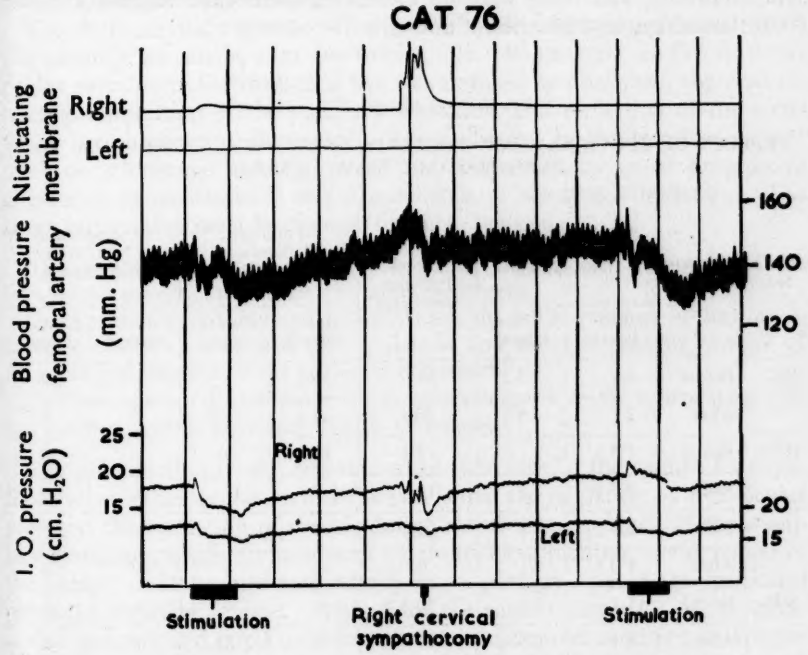

FIG. 4.—Tracing showing effect of sympathotomy on responses obtained from stimulation of a point at A 12, R 1·5, H + 6.
Note that the fall in intra-ocular pressure in the right eye has been converted into a small rise and that the response of the right nictitating membrane has been abolished. The response of the left intra-ocular pressure is unaltered.
Interval between vertical lines = 60 sec.

With regard to the response of the nictitating membrane, it can be seen that four out of the five animals showed an ipsilateral retraction which was abolished by cervical sympathotomy.

In all animals the pupillary responses were modified by ipsilateral sympathotomy; the amount of dilatation in the ipsilateral eye was reduced whilst in the contralateral eye the dilatation was unchanged or increased.

In one animal (H69, see Table) the blood vessels of the ear were observed through a microscope. During stimulation a marked vasoconstriction was noted which did not occur when the stimulation was repeated after sympathotomy.

In three animals the contralateral cervical sympathetic was divided after the effect of ipsilateral sympathotomy had been observed, but these results have not been included in the Table. The fall in intra-ocular pressure in the contralateral eye was always replaced by a rise after contralateral cervical sympathotomy. The responses of the blood pressure were again not significantly changed. The pupillary dilatations were relatively less after cervical sympathotomy, and there were no nictitating membrane responses in the contralateral eye in this series of animals.

TABLE

EFFECT OF CERVICAL SYMPATHOTOMY ON AN OCULAR RESPONSE TO HYPOTHALAMIC STIMULATION

Cat No.	Sympathotomy	Change in Intra-ocular Pressure (cm. saline)		Change in Blood Pressure (mm. Hg)	Pupillary Dilatation (mm.)		Retraction of Nictitating Membrane	
		Ipsilateral	Contralateral		Ipsilateral	Contralateral	Ipsilateral	Contralateral
H67	Before	6 ↓	5·5 ↓	20 ↓	6·5	5·5	+	0
	After	2 ↓	4·5 ↓	20 ↓	4·5	6·5	0	0
H68	Before	1·5 ↓	2 ↓	13 ↑	10	9	0	0
	After	0·5 ↑	2 ↓	16 ↑	9	10	0	0
H69	Before	2·5 ↓	2 ↓	5 ↓	6	6	+	0
	After	1·0 ↑	1 ↓	0	2	6	0	0
H73	Before	<1 ↓	3 ↑	50 ↑	9	6	+	0
	After	6 ↑	6 ↑	63 ↑	8	8	0	0
H76	Before	3 ↓	1·5 ↓	7 ↓	5	5	+	0
	After	2 ↑	1·5 ↓	5 ↓	4	6	0	0

↑ = rise ↓ = fall

Discussion

The foregoing results corroborate our previous observation that stimulation near the anterior column of the fornix frequently gives rise to a fall in intra-ocular pressure without a corresponding fall in blood pressure which, together with pupillary dilatation and retraction of the nictitating membrane, constituted an easily recognizable response-pattern. On the basis of the additional results presented here, we have widened our conception of this response-pattern and whilst regarding the fall in intra-ocular pressure as the essential feature, have recognized that the other factors (*viz.* blood pressure, nictitating membrane, and pupillary dilatation) may show considerable variation. For example, in Fig. 1 (*b*), the fall in intra-ocular pressure is accompanied by a fall in systemic blood pressure. We consider that this is a variant of the response already described, since the fall in intra-ocular pressure is too great, in our experience, to be attributed solely to the reduction of blood pressure, and the fact that a diminished fall in intra-ocular pressure occurs after sympathotomy with the same depression of blood pressure, supports this view (see H67 in Table).

The characteristic response-pattern was modified by sympathotomy in all five animals, as can be seen from the Table. In general, the fall in intra-ocular pressure in the ipsilateral eye was reduced or abolished, there was a relative diminution of the pupillary dilatation, and retraction of the nictitating membrane was eliminated. These changes in the response-pattern must be interpreted either as results of sympathotomy or as progressive alterations in the response as a consequence of repeated stimulation. The latter explanation must be rejected for three reasons:

(1) Repeated stimulation did not consistently yield weaker responses of the intra-ocular pressure, pupils, or nictitating membrane.

(2) In two animals (H68 and H76 in Table) the fall in pressure in the contralateral eye was not decreased after ipsilateral sympathotomy despite reversal of the ipsilateral response of the intra-ocular pressure.

(3) There was no diminution in the pupillary response in the contralateral eye, and in three animals there was actually an increase.

The modification of the response must therefore be the result of sympathotomy and the conclusion to be drawn is that the reduction of intra-ocular pressure, the retraction of the nictitating membrane, and part of the pupillary dilatation, which are produced by electrical stimulation in the region of the fornix, must be mediated wholly or in part by way of the ipsilateral cervical sympathetic nerve. Also, since the similar responses of the intra-ocular pressure and pupil on the contralateral side were modified by sympathotomy on that side, it follows that each (unilateral) centre exercises some bilateral control.

In the introduction we mentioned the possibility that the response of the intra-ocular pressure to stimulation in this area of the hypothalamus results

from a constriction of the ocular blood vessels mediated by the cervical sympathetic nerve. It is true that the actual changes in intra-ocular pressure provide only indirect evidence of vasoconstriction, but its occurrence is strongly suggested by the observation of an associated vasoconstriction in the ear. That this reaction may not be confined to the eye is to be expected (see Gloster and Greaves, 1956, 1957).

The effect of cervical sympathotomy upon ocular responses to diencephalic stimulation has also been examined by von Sallmann, Macri, Wanko, and Grimes (1956), who reported that retraction of the nictitating membrane was abolished by sympathotomy, whereas pupillary dilatation was unaffected. Their findings with regard to the response of the intra-ocular pressure are not comparable with ours, since they stimulated a region which produced simultaneous *rises* of *both* blood pressure and intra-ocular pressure, neither of these being modified by sympathotomy. With regard to pupillary responses, Weinstein and Bender (1941) showed that the dilatation resulting from stimulation of the ventral hypothalamus of the cat was reduced by cervical sympathotomy, thus indicating that the response was mediated in part by this nerve. Ranson and Magoun (1933) drew attention to the fact that, although pupillary dilatation could be obtained by stimulation in many parts of the hypothalamus, one of the main regions yielding this response was situated around the fornix. Hess (1954) also reported maximal pupillary dilatation on stimulation of the so-called nucleus perifornicalis, an ill-defined area which has been described by Rioch (1929) and by Ingram, Hannett, and Ranson (1932). The area from which we obtained the characteristic response-pattern of a fall in intra-ocular pressure, pupillary dilatation, and retraction of the nictitating membrane appears to conform anatomically with the region described by these workers, and we regard our results as an extension of previous findings which have been limited to observations on the pupils. Although our results establish that stimulation in this area of the hypothalamus activates the sympathetic nerve supply to the eye, it is clear that the occurrence of simultaneous changes in the blood vessels of the ear and in the systemic blood pressure show that further evidence is necessary in order to determine whether any part of this area is associated *exclusively* with the eye.

Summary

(1) Stimulation in the region of the anterior column of the fornix of the cat produces a typical response characterized by:

(*a*) a bilateral fall in intra-ocular pressure;
(*b*) pupillary dilatation;
(*c*) occasionally retraction of the nictitating membrane;
(*d*) usually a rise of systemic blood pressure.

(2) This response is modified by cervical sympathotomy as follows:

(*a*) the fall in intra-ocular pressure is abolished;
(*b*) the pupillary dilatation is reduced;
(*c*) retraction of the nictitating membrane is abolished;
(*d*) the blood-pressure response is not significantly affected.

(3) This indicates that the response is mediated by way of the cervical sympathetic nerve, and that, as this response is bilateral, stimulation on one side of the hypothalamus results in a discharge in the cervical sympathetic nerve trunks on both sides.

(4) The response is not confined to the eye since simultaneous constriction of the vessels of the ear has been observed.

We should like to thank Sir Stewart Duke-Elder for his continued help and advice; also the Alexander Pigott Wernher Research Trust for a grant covering the expenses of this study.

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OPHTHALMIC DISEASE IN HONG KONG*

BY

G. C. DANSEY-BROWNING

Hong Kong

HONG KONG, which lies on the south coast of China just within the tropics, consists of some 193 islands surrounding a mountainous peninsula to the east of the Pearl River. The capital, Victoria, situated upon the island of Hong Kong (Haven of Fragrant Streams), faces the Kowloon (Nine Dragons) peninsula across a perfect natural harbour. At the present, the Colony's population ranges upwards from 2½ million, 98 per cent being Chinese. Some 2 million persons live crammed into the 12 square miles of the urban areas of Kowloon or Victoria, giving in the slum areas a density of 2,000 per acre (Annual Departmental Report of the Director of Medical and Health Services, 1954–56). This multitude due to immigration and a prodigious birth-rate shows an annual increase of 80,000 (Annual Report, 1956). Only 50 of the remaining 380 square miles of the Colony's area are suitable for agriculture, or indeed for any other purpose. Yet some further half million persons make their living either by farming or by fishing the seas surrounding Hong Kong.

ECONOMY

Established some 116 years ago, Hong Kong quickly became one of the principal *entrepôts* of the China Trade—a position retained until 1949. Recent political changes on the mainland of China have resulted in a shift to light industry with emphasis on textile and plastic manufactures. Also the Colony has had to absorb nearly one million refugees in search of security and employment. The majority of the Chinese population hail from the neighbouring provinces of Kwangtung and Fukien. They are only one generation removed from an agricultural society based on fixed traditions, and operating under rules that scarcely have changed in 2,000 years. In consequence, these Cantonese—a generic term which embraces Hong Kong's Chinese—have a completely open mind as to the relative values of the Oriental and Occidental systems of medicine. On the whole they tend to prefer at first instance the ministrations of their own herbalists. But once diseased, being a true race of shopkeepers, they are perfectly willing to visit all available practitioners of either system, "shopping around", eventually to choose the one who will promise a certain cure at the cheapest fee. In

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ophthalmology, at any rate in the poorest quarters, the balance at the moment is nicely adjusted between the obvious value of western antibiotics and the excellent if erratic knowledge of herbs and simples that the indigenous practitioners have acquired through generations of trial and error at their patients' expense.

Incidence of Disease

In 1953, estimates of the incidence of ophthalmic disease amongst the very poor in the Colony indicated a potential attendance of 150,000 per annum. The small white-collar class were adequately catered for by some fourteen private practitioners of ophthalmology, but their fees were such that a coolie on a basic wage of 3 to 4 $ a day could never afford to attend their clinics. The Free Hospitals of the Chinese (Tung Wah) Group could offer the poor a choice of either Western or Eastern medicine, but their three part-time ophthalmologists could only deal sketchily with 30,000 attendances per annum, and there were definite limits to the cost of drugs and treatment provided. It was therefore inevitable that the small government ophthalmic service should need considerable expansion as soon as an effective system of out-patient records had been established and certain surveys of ophthalmic disease had been carried out.

The 1953 surveys showed that the principal diseases presenting at the urban clinics were as follows:

(A) Acute infection of the anterior segment of the globe (ophthalmia), associated with varying degrees of subnutrition.
(B) Phlyctenular disease.
(C) Syphilis.
(D) Trachoma.
(E) Untreated cataract.
(F) Glaucoma.

Further factors affecting the incidence of disease were also disclosed by these surveys:

(1) *Overcrowding*.—In the slums of Wanchai or Shamshuipo, the area of bed space that an adult can acquire is only 15 sq. ft—all too often this is not only a place to sleep, but also the space wherein he has to earn his livelihood. In consequence not merely is the epidemic spread of ophthalmia accelerated, but the high racial tuberculosis morbidity is aggravated; although on the whole the Chinese show a low susceptibility to this disease. Hong Kong's weekly notification of deaths from tuberculosis averages well over 45, while the freshly diagnosed cases of the disease are higher than 200 per week. Some 78 per cent. of deaths in the male age group 40–44 years, and 30 per cent. of deaths in infants under 5 years of age are due to tuberculosis (three-quarters of the latter being due to tuberculous meningitis).

In 1954 a series of 440 children with phlyctenular disease aged 8 years or under was tuberculin-tested. Some 85·9 per cent. of these were reported as "T-T positive". All the children in the series had their chests *x*-rayed, with immediate

reports in 48 cases of active pulmonary tuberculosis. Of the whole, 20·5 per cent. were reported either as being actively tuberculous or else as showing radiological stigmata significant of this disease.

(2) *Diet.*—The key to the whole problem of effective mass treatment of ophthalmic disease, not merely in Hong Kong but throughout South East Asia, lies in the dietary idiosyncrasies of the inhabitants. During the first year of life, the Cantonese infant is kept at the breast, and a recent survey shows that during this period the weight-growth rates closely parallel those for infants of other races. However, directly the Chinese infant is weaned the weight-growth rates slow up. In Hong Kong, when artificial feeding has to be resorted to, the economics of poverty bring it about that infants receive only unfortified tinned milk or a rice-mush known as *congee*. The Cantonese dislike bread and avoid all forms of dairy-produce in their diet. Throughout the year the poor exist upon bowls of polished rice garnished with a few vegetables and a little salt-fish, except for the traditional feast of pork or chicken at the Lunar New Year. The majority of our series of phlyctenular disease showed a heavy infestation with *Clonorchis sinensis*; this trematode has as an intermediary host the fresh-water carp—a staple of the Cantonese diet—and it is held by some authorities to affect over 65 per cent. of the population of Kwangtung.

The school health scheme operating in the Colony reports that 31·8 per cent. of its entrants are found to be of poor or subnormal nutrition, and that a further 20·6 per cent. are noted at this time as border-line cases meriting further observation. 18·2 per cent. of these entrants required immediate remedy of physical defects—the ophthalmic service providing spectacles for approximately 10 per cent. of the whole entry.

Dietary factors are responsible for the lack of resistance to ocular infection amongst our patients, reaching in extreme cases to pure keratomalacia, and assisting in the production of a high incidence of phlyctenular disease. In 1955, an analysis of 300 children on their first attendance at our urban clinics showed 44·6 per cent. to be suffering from ophthalmia/subnutrition and a further 17·0 per cent. from phlyctenular disease. Like the English the Chinese believe that to be "good" all medicine must be "nasty or searching". In consequence, the salves and powders that the local necromancers, herbalists, acupuncturers, and the like instil into the eye are in all essentials abrasive, and the excoriations so produced are all too often difficult to differentiate from massive crops of phlyctenules.

(3) *Syphilis.*—The Chinese claim that this disease is purely a "foreign-devil" importation of the last 150 years. Be that as it may—in 1954 some 6 per cent. of all pregnant women in Hong Kong were found to be suffering from unsuspected syphilis, and in the same year an analysis of blindness in males showed serologically that 10 per cent. were due to syphilis. The ophthalmic clinics that year referred some eighty cases of proven syphilitic optic atrophy to the Social Hygiene Clinics —not one of which had previously received any form of anti-luetic treatment. Curiously enough, the Cantonese are convinced that blindness is the inevitable result of the sins of the sufferer, and the above findings may be held to give support to this belief; this has been one of our principal stumbling blocks in the struggle to enlist public support for blind-welfare.

(4) *Trachoma.*—Hong Kong has always had an inadequate water supply for its large community, and in the tropics this poses many public health problems, not the least of which is the facility with which communicable eye-disease spread. To aggravate matters, the Chinese, whilst fastidious in their personal hygiene, are completely oblivious of the necessity for communal measures. Conservancy is regarded as a western-fad; expectoration at all times and in all places is a general habit; the communal face towel wrung out in tepid water is used by all the company attending eating houses and places of entertainment to refresh the face and hands. Our urban clinics in consequence show a constant 15–17 per cent. attendance for trachoma, but the incidence is considerably higher in the rural clinics held for farmers and fisherfolk for whom hygiene does not exist. The children attending the middle-class schools show very little trachoma, but in the orphanages, rural schools, and institutions, the incidence amongst the inmates is about 11 per cent.

(5) *Less Common Diseases*

(*a*) Peculiar to this region and held by some authorities to be bound up with the hot humid climate is the Cantonese disease. Each year the ophthalmic clinics expect some fifty or more patients in early middle age to report with a paralytic strabismus or other ophthalmic signs that on investigation prove to be a hitherto unsuspected but by now intractable neoplasm: naso-pharyngeal carcinoma.

(*b*) Leprosy in Hong Kong provides 11,000 annual attendances in the government clinics, approximately 500 new cases being diagnosed each year. In the main, the ophthalmic signs are either paresis affecting the orbicularis oculi or uveitis in the tuberculoid type of Hansen's disease. We have only seen one case of the classical lepromatous pannus in the last 4 years, but a retino-choroiditis the initial appearance of which is extremely difficult to differentiate from diabetic retinopathy is frequently seen.

(*c*) The Chinese in Hong Kong are firm believers in vaccination (red-bean-sowing) and in consequence the amount of blindness due to smallpox (the heavenly red-bean flower disease) is negligible. However, after the acute infectious fevers and in particular after measles, tradition has it that the child should not receive a full diet for one month, and such patients are all too often rescued just upon the brink of keratomalacia.

Public Health Measures

Preliminary surveys of the Colony's blind population indicated that nearly 50 per cent. of these unfortunates claimed that they had lost their sight before the age of 10 years. United Kingdom surveys state that only 6·2 per cent. of blind persons are so disabled before the age of 15 years, and 82·3 per cent. are reported to have lost their sight after the age of 50 (Sorsby, 1953). In 1954, the government eye clinics of Hong Kong freed all children of the age of 10 and under from any charges for treatment, drugs, hospitalization, etc. Otherwise a fee-for-service is charged of $1 per patient. The overall value of this to a family of seven or ten children existing on a coolie's pay cannot be exaggerated. Immediately, instead of seeing children in the last

stages of keratomalacia, we were able to treat acute infections of the eye in the earliest stages. By the middle of 1953, all government clinics, closely copied by outside practitioners, started to treat trachoma with Aureomycin/Terramycin/sulphonamide therapy under strict W.H.O. classification and discipline. At the same time, in all cases of disease in children, we reinforced topical therapy with systemic antibiotics and nutritional supplements (mainly vitamins A and D) to get rapid initial response to treatment. The results have been most gratifying, and now we boast that we only get "blind babies" from Kwangtung, and we know that our "yellow trachoma ointment" has a wide reputation throughout South China. Unfortunately Hong Kong's prolonged therapeutic honeymoon with the antibiotics shows signs of being on the wane, and there is also a noticeable increase in the photo-sensitive reactions to sulphonamide therapy similar to that now occurring in the Mediterranean area.

All too often in ophthalmology, the role of welfare as a most vital weapon in our armamentarium is forgotten. The ophthalmic almoners here have to deal with approximately one in 25 of all our fresh cases; they achieve the most excellent results, giving advice on economic problems. Gifts of food parcels and home-visits to deal with problem parents help to reduce the numbers of our "chronics" and speed the pace of cure.

Blindness

Mackenzie and Flowers (1947) estimated that there were nearly 2 million blind persons in China. Such figures may seem quite fantastic, but there can be no doubt that the numbers must be astronomical in a population recently stated to approach 600 million persons.

We base our estimates of Hong Kong's blind population upon United Kingdom surveys, which would indicate that the number of persons with visual acuity of hand movements, perception of light, or less would be about 3,000. Some 551 of these were surveyed in 1953 (Table I).

TABLE I
MAJOR CAUSES OF OPHTHALMIC DISEASE

Cause	Percentage of Cases
Ophthalmia/Subnutrition	44
Untreated Cataracts	16
Trachoma	11
Injury	10
Syphilis	5
Congenital or Hereditary Disease	5
Glaucoma	3·5
Uveitis or Optic Atrophy of Unknown Aetiology	2·5
Smallpox	1
Leprosy	1
Degenerative Disease	1
Total	100

This preliminary survey, whilst leaving unanswered many questions, clearly emphasized the fact that three-quarters of the blindness in Hong Kong was preventable. The major discrepancies with the United Kingdom figures lay in the different ages at onset, and in the figures for untreated cataract and glaucoma (30·4 and 14 per cent. respectively in the United Kingdom; Sorsby, 1953).

In the survey of 800 blind adults, summarized in Table II, the incidence of cataract and glaucoma in Chinese and European adults clearly approximated. The characteristics of male blindness in Hong Kong appear to be the high incidence of syphilis (as against 1 per cent. in the United Kingdom; Sorsby, 1953), and of injuries consequent on nearly 50 years of civil war, flood, and famine. In Hong Kong, as indeed throughout the Orient, there is no idea that heavy work is unsuitable for women, hence the parity between the sexes in "peace-time" traumatic causes of blindness. Furthermore, the Colony is only lightly industrialized, so that the summation of all types of trauma does not approach the 24·7 per cent. of traumatic blindness encountered in the figures for the heavily industrialized United Kingdom. In fact, the textile and plastic factories of Hong Kong are generally of recent construction, rivalling the best in other countries in layout, equipment, and employee-welfare services. Yet side by side with these exist concerns in which the employees live and work in conditions and under an apprenticeship system analogous with those of Tudor London.

TABLE II

ANALYSIS OF 800 BLIND ADULTS IN 1954

Ophthalmic Cause			Percentage of Cases			
			Female		Male	
Major (More than One per cent.)	Ophthalmia/Subnutrition		29		24	
	Cataracts	Simple	25·0	30·4	12·0	10
		Complicated	5·4		8·0	
	Trachoma		14·5		9	
	Glaucoma	Simplex or Narrow Angle	8·0	9·5	5·0	5·7
		Secondary	1·5		0·7	
	Injury	Peace-time	4·5	4·6	4·0	17
		War-time	0·1		13·0	
	Syphilis	Congenital	0·1	2·6	0·2	10·2
		Acquired	2·5		10·0	
	Non-specific Uveitis		2		3·5	
	Non-specific Optic Atrophy		1		1	
All Other Minor Causes (Less than One per cent.)			100		100	

Nearly 90 per cent. of all adults who had lost their sight because of ophthalmia/subnutrition claimed that this had occurred before the age of 10 years.

The survey of 200 blind children summarized in Table III, reflects the points noted elsewhere regarding the age at onset of blindness, and the high birth-rate. Tuberculous meningitis and epidemic encephalitis are in the main responsible for the figures for optic atrophy.

Most of the children surveyed came from institutions in the Colony, the slight bias towards females being due to the fact that the female child is of little economic value to a Chinese family, and in consequence readily discarded in times of disease and stress. One of the more interesting causes of uniocular blindness in children cannot feature in this Table. At the period of the Chinese New Year, Hong Kong is ablaze for 48 hrs with the explosion of fireworks, and we normally expect to see fourteen to twenty cases of grave ocular damage amongst children. Fortunately not many eyes progress to enucleation, but it is of interest that the casualty figures at this season approach those in London on the evening of Guy Fawkes Day.

TABLE III
ANALYSIS OF 200 BLIND CHILDREN IN 1954

Ophthalmic Cause			Percentage of Cases	
			Female	Male
Major (More than One per cent.)	Ophthalmia/Subnutrition		71	60
	Congenital Defects	Cataracts	12	9·5
		Other than Cataracts	10 } 22	8 } 17·5
	Non-specific Optic Atrophy		5	6
All Other Minor Causes (Less than One per cent.) ..			100	100

In 1955, the case histories of some 1,071 blind persons were examined to find out how much blindness was due to a combination of several diseases, and 154 were found to come into this category.

Amongst eighty females, glaucoma with secondary cataract (10), trachoma with senile cataract (9), senile cataract with the other eye blind from peace-time trauma (9), non-specific uveitis with subsequent cataracts (9), and senile cataracts with secondary glaucoma (8), were the main combinations.

The 74 males on the other hand presented non-specific uveitis with subsequent cataract (11), acquired syphilis with subsequent cataract (7), glaucoma with subsequent cataract (7), and senile cataract with the other eye blinded by peace-time trauma (7) as the main combinations.

Ophthalmic Arrangements

The government eye clinics have now expanded rapidly to deal with over 85,000 attendance in 1956, and the figures reached 100,000 during 1957. Two

large well-equipped ophthalmic centres cater for the urban areas, and from them a staff of twenty, including five ophthalmologists, provides 28 clinic-sessions monthly at eight out-stations to cover the whole of the Colony. Our beds are only ten in number, with a turn-over of 32 patients each during the year, but the majority of our major and intermediate operations (2,500–2,900 cases per annum) is carried out as "office-surgery" at the major centres. The patient is securely stitched and sent home after operation, and we feel that the complication rate appears to be no greater than in those who were fully hospitalized. Over 75 per cent. of our cataract surgery is perforce carried out as "office-surgery".

Summary

Surveys of ophthalmic disease and of blind persons in Hong Kong are described. The expansion of the government ophthalmic service as a result of these surveys shows the development of the role of preventive ophthalmology.

My thanks are due to the Hon. Director of Medical and Health Services, Hong Kong, for permission to publish this article, and to my colleagues in the Ophthalmic, Health, Tuberculosis, and Social Hygiene Services for their constant help.

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PROTECTION OF THE GRAFT WITH CONDOM IN KERATOPLASTY*

BY

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IN the course of time several more or less successful procedures have been recommended to protect corneal grafts. The fact, however, that ophthalmic surgeons with many patients recommend different methods proves that a method of choice still does not exist. The ever-increasing number of keratoplasty operations shows, however, that the problem has passed the stage of experimentation, and that the operation may now be regarded as a routine procedure. A simple and effective method of protecting the graft has thus become an important topic.

Vannas (1939) recommended a small oblong rubber membrane for the protection of the transplant. He fixed the covering rubber near the limbus below and above with three to five episcleral sutures. He emphasizes that condom is the best covering material because of its thinness, elasticity, and transparency. This simple and easy method has not been universally adopted, chiefly because the extensibility of the oblong rubber membrane has not proved entirely satisfactory.

We have been performing the great majority of our keratoplasties for more than 10 years by the method recommended by Vannas, and have had ample opportunity to discover all its difficulties. The original method of fixation is not quite satisfactory because of the looseness of the longer sides of the oblong membrane, but the general idea is excellent and with slight modifications this may be regarded as the safest means of indirect protection. The covering should be made circular, slightly larger than the host cornea, and fixed in several places near the limbus to the episclera.

Harms (1948) modified the method of Vannas in much the same way, but his article found no response in the non-German literature, partly because it was very brief and partly because he solved only part of the problem. Even with his modification some technical difficulties remained, and the fixation of the rubber membrane was a rather fiddling and difficult job.

The following procedure is an improvement on the Vannas-Harms condom method. A rubber membrane, usually with a radius 2 mm. greater than that of the host cornea, is cut with a suitably sized punch of the type used for punching leather. The condom membrane is put on a soft wooden

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plate, and 1·5 mm. inside its margin fine sewing needles are used to insert 6/0 black silk at 12, 2, 4, 6, 8, and 10 o'clock. To prevent the folding of the condom membrane the inserted threads are tied over fine pincers. The covering rubber membrane is then perforated either with a fine platinum cautery or with an Elliott trephine 1 mm. in diameter. The loop of the cautery is made white hot and the size of the holes is regulated by the perforation time. These perforations are made in a circle 3 mm. inside the margin of the membrane, their number depending on the diameter of the protecting membrane. Besides the marginal perforations a hole is also made in the centre.

The condom membrane is sterilized for 24 hrs in a penicillin bath (10,000 I.U./ml.) or for 3 minutes in boiling water. After sterilization the rubber membrane is placed in sterile gauze, and the dried threads are arranged ready for use.

After exposure with a Pretori speculum, bridle sutures are inserted into the superior and inferior rectus muscles to fix the globe and make the insertion easier.

The rubber membrane prepared beforehand is then centred on the host cornea, the sutures lying at 12, 2, 4, 6, 8, and 10 o'clock (Fig. 1).

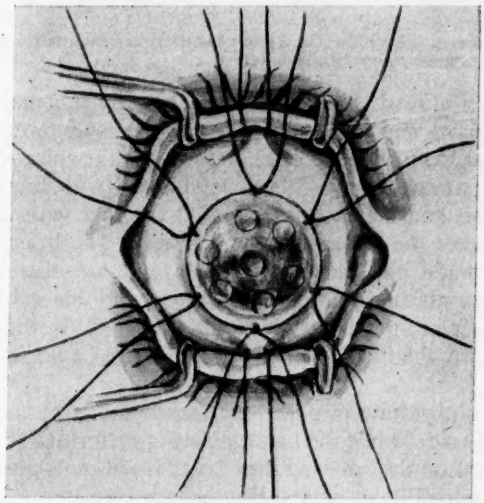

FIG. 1.—The condom membrane prepared beforehand is placed on the host cornea. The threads are at 12, 2, 4, 6, 8, and 10 o'clock, and the central hole is at the centre of the host cornea.

The thread at 6 o'clock is inserted 2 mm. below the limbus into the episclera, knotted immediately, cut short. The episcleral insertion of the needle can be facilitated by first making small superficial incisions into the

sclera with a knife or a keratome. The threads at 4 and 8 o'clock are then inserted in the same way (Fig. 2).

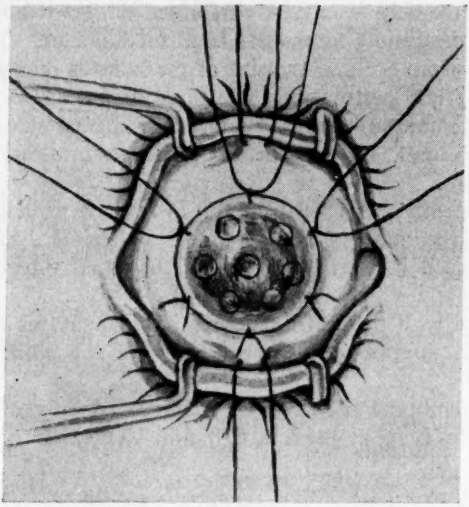

FIG. 2.—Position after the three lower episcleral threads have been inserted, tied, and cut.

When the lower threads have been tied, the rubber membrane does not slide about any more and no further difficulty is encountered in fixing it.

The surgeon inserts the thread at 12 o'clock into the episclera, the assistant helping him by stretching the rubber membrane with fine forceps. This thread is not tied, and the assistant holds it together with the upper bridle suture which makes the other insertions easier. The insertions at 10 and 12 o'clock are made like the previous one and are also left untied. In anchoring the membrane care must be taken that the rubber is properly stretched; the protecting membrane should lie smoothly without folding on the host cornea, as the fixation effect required can be achieved only in this way.

When all the sutures are in place, the membrane is turned down and the threads are held aside, while the keratoplasty is performed (Fig. 3, opposite).

After the operation the covering membrane is put in its place and the three last sutures are tied (Fig. 4, opposite).

The protecting membrane is usually removed on the 8th day. In our experience the sutures of the rubber membrane will not tear out if it is properly stretched and the tension is satisfactory.

Summary

Condom, as recommended by Vannas (1939, 1950), is the best material

for the indirect protection of a corneal graft. His method was modified by

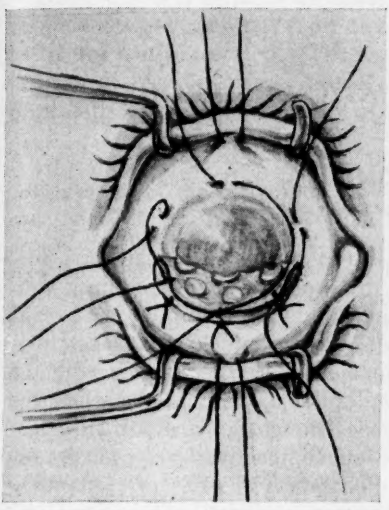

FIG. 3.—The threads at 10, 12, and 2 o'clock have been inserted and the rubber membrane has been turned back.

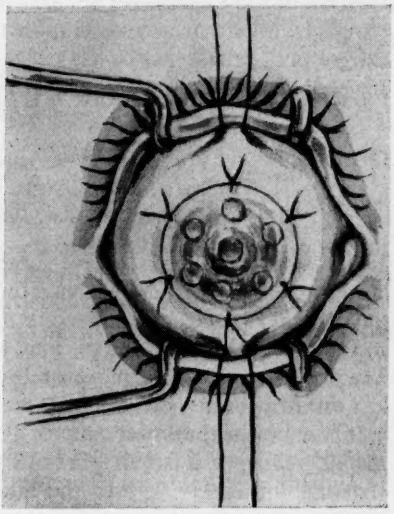

FIG. 4.—Position of the rubber membrane after grafting has been performed and the sutures have been tied.

Harms (1948, 1956), who cut a circular rubber membrane and sutured it to the episclera at six points on the limbus.

This method is a further development of the Vannas-Harms condom method, which simplifies the performance of the operation, and holds the transplant perfectly in place.

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REPLACEMENT OF TRAUMATIC IRIS PROLAPSE*

BY

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THE various text-books of eye-surgery, in dealing with traumatic iris prolapse, give the traditional advice to cut the prolapsed iris tissue flush with the corneal surface, after having firmly pulled out the iris through the wound so that the cut edges of the iris may retract away from the corneal laceration. The result is usually a more or less broad disfiguring coloboma with the known drawbacks of dazzling and photophobia, particularly when the coloboma is in the lower part of the eye. Though Stallard (1950) recommends reducing a small post-operative subconjunctival iris prolapse by stroking the iris back into the anterior chamber, in cases of penetrating injuries where the iris is in direct communication with the conjunctival sac and infection is likely, the principle of his treatment is not replacement but abscission, making as small a colomba as is compatible with avoiding anterior synechiae.

But even with the excision of a prolapse performed in the classical way one may not always achieve an ideal retraction of the pillars away from the wound, especially where the wound is concentric to the limbus and about midway between limbus and corneal centre and/or has slanting edges. Pulling the iris sufficiently out in such cases is dangerous in that the iris may tear at its root. Spaeth (1948) recommends a fresh keratome incision at the limbus near the perforation and withdrawing the iris from the perforation wound through this new incision. For some of these cases, particularly in freshly sustained injuries, he advises releasing the iris from the perforation and smoothing it flat. He states that, in general, these cases of traumatic iris prolapse are best handled by abscission of the prolapsed iris.

Duke-Elder (1954) writes that prolapsed uveal tissue should never be replaced because of the risk of introducing infection into the eye.

Because the new antibiotics now make it possible to prevent or combat nearly all forms of infection, and because of the beneficial effect of the corticosteroids on inflammatory reactions, the excision of an iris prolapse seems to us no longer necessary. On the basis of experience gained from handling anterior synechiae in leucoma adherens prior to corneal grafting, a method has been developed of replacing prolapsed iris by a manoeuvre from inside

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the eye, thus avoiding the traumatizing effects of trying to reinsert the iris with a spatula from outside through the corneal wound.

This method is suitable for cases in which the prolapse is not more than about 4 days old and not more than 6 to 7 mm. in extent, and in which the iris is not badly lacerated. The results have been so satisfactory that we feel justified in recommending it as a standard procedure for the management of prolapsed iris in the majority of suitable cases.

Technique

The extent of the perforation, its form and course, the amount of prolapsed iris, the nature of the incarceration, the degree of mutilation of the iris, the presence of foreign material in the wound or anterior chamber, and the risk of eventual injury to the lens all have to be investigated.

A smear and a culture is taken from the conjunctival sac to ascertain whether infection is present and to which antibiotics the bacteria are susceptible. Intensive local and systemic treatment with antibiotics is started immediately to achieve conditions as aseptic as possible up to the moment of operation which should not be unduly delayed.

The preparation of the patient and of the eye are the same as for any other intra-ocular operation: general and local analgesia and akinesia, retrobulbar injection of Novocain with adrenaline and hyaluronidase (this also in the case of general anaesthesia), lateral canthotomy if necessary, and bridle or traction sutures at the limbus. In badly lacerated corneal wounds with irregular and gaping edges with little chance of an exact apposition by intracorneal sutures, an adequate conjunctival flap is prepared. If the prolapse is covered with fibrin or by a layer of regenerated epithelium as in delayed cases, this is removed by gentle irrigation with warm saline, the floating tags being carefully picked up with a fine forceps. Adhesions between iris and wound edges are broken by carefully introducing the tip of a fine repositor.

The anterior chamber is then opened by a slanting trap incision at the limbus (Fig. 1).

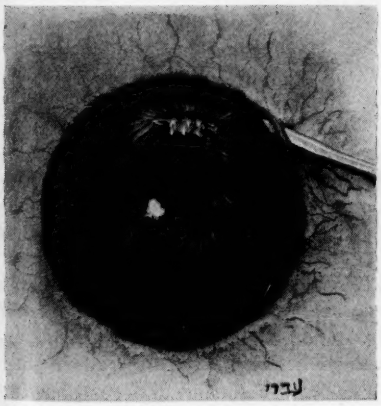

FIG. 1.—The anterior chamber is opened by a trap incision with a cataract knife at a distance of 4 to 6 mm. from the prolapse.

The position of this opening depends on the site of the iris prolapse and should be situated about 5 to 7 mm. from the prolapse so that a spatula introduced into the chamber lies between the limbus and the corneal wound and as tangentially to the wound as possible. For this incision a narrow cataract knife is used instead of a keratome; the knife makes an incision even into a completely soft eye with ease and safety. A good knife-needle may be used. There are, as a rule, no difficulties even if the chamber is not present, so long as the entrance of the tip of the knife into the chamber is carefully watched and the knife is guided along the anterior chamber angle between iris and cornea until the inner opening is about 1 mm. wide.

Through the incision a narrow, rounded cyclodialysis-spatula is introduced and guided carefully between iris and cornea so that it lies halfway between the limbus and the iris prolapse, its tip reaching beyond the distal margin of the prolapsed iris (Fig. 2). By a gentle sweep of the spatula from the periphery towards the centre, the prolapsed iris is swept back into the chamber and smoothed out. In broad prolapses or adhesions between iris and wound edges the lips of the corneal wound may at the same time be made to gape by spreading them with a second spatula. In extensive prolapses it is not always possible to replace the prolapse by one sweep. In these cases the reposition is performed step by step, starting with the part which is nearest to the limbal incision, or replacing an unmanageable portion from another better placed incision.

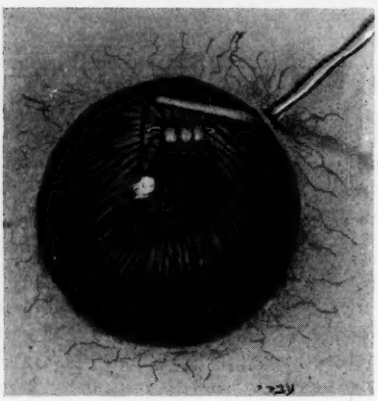

FIG. 2.—The iris prolapse is replaced by a cyclodialysis spatula introduced into the chamber through the trap incision.

After the replacement, as many intracorneal sutures are inserted as are necessary to accomplish an exact apposition and an air-tight closure. If exudate has already formed in the anterior chamber or if hyphaema is present, this is washed out, the tip of the irrigator being introduced between the wound edges. The corneal sutures are then tied.

The anterior chamber is restored by injection of sterile air through the trap incision. If the air escapes through the corneal wound an additional corneal suture is placed at the site of the leakage. When a conjunctival flap has been prepared, it is drawn into position, so preventing any escape of air from an irregular corneal wound which could not be exactly closed. Penicillin and cortisone are

injected subconjunctivally, atropine and Polyfax ointment are applied to the lower fornix, and both eyes are bandaged for 2 days.

In iris prolapses at the limbus or through the sclera adjacent to the limbus, though one may try to replace the iris by the method used when the iris prolapses during an intra-ocular operation, an exact reposition through a small wound without any entanglement of iris strands may be very difficult, even after relieving the herniated iris from the pressure of the aqueous by a peripheral iridectomy. It is much safer to apply the procedure described above, with the small modification that the incision is made into the sclera down to the ciliary body as for cyclodialysis, about 4 to 5 mm. from the limbus and parallel or perpendicular to it. Through this incision a cyclodialysis spatula is introduced between sclera and ciliary body up to a point behind the prolapse, and both the ciliary body when prolapsed and the iris are replaced by a sweeping movement towards the chamber (Fig. 3). Through the same opening air is injected with a special cannula into the chamber, after the corneo-scleral wound has been sutured.

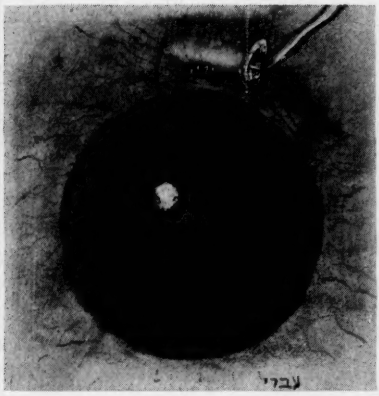

FIG. 3.—Replacement of an iris prolapse at the corneo-scleral junction through a scleral incision in the manner of a cyclodialysis operation.

In large corneal wounds with extensive prolapse of lacerated iris, it is not right to replace the iris if it is hanging outside in ribbons. A neat coloboma will always be safer than rags of iris inside the chamber. But in these cases it is still unnecessary to excise the whole prolapsed iris. Only the lacerated parts should be excised, and the intact portions of the iris may be replaced as described above.

If the iris prolapse is complicated by a simultaneous traumatic cataract, one should proceed as though there were no cataract if there is only a small wound in the anterior lens capsule with a circumscribed opacity. There is more chance that the wound in the capsule will be closed and progression of the cataract stopped when the iris is replaced and is covering the opening in the capsule than when the iris is excised and the capsule wound exposed to the influence of the aqueous. In cases of wide lacerations of the lens capsule and quick opacification of the lens, the surgeon must decide whether to excise the iris or replace it. We prefer to replace a small to moderate prolapse, and after replacement as much as possible of the anterior capsule is removed through the wound. The anterior chamber is then thoroughly irrigated and larger pieces of lens cortex and nucleus

removed by means of a small Jaeger's spoon. These manipulations are started after corneal sutures have been placed. In some of these cases it may be preferable to delay the surgical repair for one or two days and to utilize the delay for intensive treatment with antibiotics; the postponement has the advantage that the lens becomes more opaque and disintegrates so that its removal is easier and more complete.

Results

In the last 3 years 42 cases of perforating injuries of the cornea complicated by a simultaneous iris prolapse have been admitted to the Eye Department of the Government Hospital at Tel-Hashomer, not including cases of severe lacerations and ruptures of the eyeball due to road accidents, mine explosions, and battle injuries. From these 42 cases twenty were selected as suitable for a replacement of the prolapsed iris. In the majority the prolapse was replaced 24 to 36 hrs after the accident, but in four cases 2 days had passed and in two cases 3 days. Even in these delayed cases no special difficulties were encountered in replacing the iris after having freed it from its adhesions to the corneal wound and stripped the fibrin from its surface.

In twelve cases the corneal laceration was not more than 4 mm. long, in four cases it was 5 to 6 mm. and in the remainder about 7 mm. In one exceptional case in which the replacement was successful, the corneal wound and the iris prolapse extended from limbus to limbus 2 mm. below the corneal centre.

In most cases the wound was more or less linear, but in three it was irregular, bruised, and gaping with loss of corneal tissue.

In five cases a traumatic cataract was present. In three of these most of the lens matter and a large piece of the anterior capsule were removed at the same session, in two the cataract was removed later, and in one it was left *in situ* with a visual acuity of 6/24.

In two cases the wound was situated in the sclera and the replacement of the incarcerated iris was performed through an incision in the sclera as described above.

Complications.—In one case an implantation cyst had developed, and this was later removed successfully by a basal iridectomy. In about half the cases small posterior synechiae developed at the site of the prolapsed sphincter without greatly disturbing the round shape of the pupil. In some cases small patches of iris atrophy confined to the prolapsed part appeared, and in these parts the normal relief of the iris was lost. No other complications, no infections or serious inflammations and no traumatic cataract which could be related to the operative procedure were encountered.

Comment

Two decisions have to be made in considering the repair of a traumatic iris prolapse: whether to excise or to replace the prolapsed iris, and what method of replacement to use.

The chief argument against replacement of a traumatic iris prolapse has been the fear of introducing infectious material into the eye with the iris that has been exposed. Our experience, though limited to only twenty cases, suggests that this risk is no greater than in any other intra-ocular operation when adequate prophylactic measures are taken. In none of the operated cases did infection develop after the reposition and no additional inflammatory reaction was observed. The only complications seen were an implantation cyst in one case (the implantation having possibly occurred at the time of the original injury), and insignificant small posterior synechiae with atrophy of the adjacent iris at the site of reposition in about half the cases.

The results of this method whereby the prolapsed and incarcerated iris is freed from the corneal wound by a manoeuvre from inside the chamber have been so satisfactory that it might be used as a standard procedure where replacement of an iris prolapse is considered.

As to the general indications for replacement of iris tissue that prolapses after laceration, Callahan (1956) has reached the same conclusions. Because of the resulting photophobia, iris deformity, and occasional reduction of vision, he prefers excision to replacement of a small to moderate prolapse, and hopes that this will gradually become the standard method of management. The fundamental difference between his technique and ours lies in the method of replacement. Callahan reinserts the iris through the laceration with a spatula or iris hook. Everybody who has tried this manoeuvre knows how difficult, even impossible, it can be, because the prolapsed and incarcerated iris, being friable and without tonus, tends to come out again and again, even after the aqueous has been drained. This method of reposition is more difficult the smaller and more bevelled the corneal wound is, and it is almost impossible to perform it in an exact and gentle way when the direction of the bevel is towards the periphery and the wound near to the limbus. In wounds bevelled towards the centre there is the additional danger of injuring the lens with the spatula when one is trying to free the unyielding iris from the posterior lips of the wound. In both eventualities the manoeuvre of direct reposition through the wound is apt to damage the iris still more. No such difficulties are encountered when the iris is swept back into the chamber by the gentle pull of the spatula stretching the iris between its root and the point of engagement. In this manoeuvre the iris is not so much reinserted as replaced.

A second minor difference between the two techniques is the sequence of the operative steps. Callahan inserts the intracorneal sutures as an initial step in the repair, because in his opinion there is less danger of injuring the lens if it is protected by a cushion of aqueous which escapes when the iris prolapse is excised or replaced beforehand. We have never found that there is a special danger of injuring the lens when placing intracorneal sutures, even when there is no anterior chamber at all, even under the more difficult

conditions of suturing penetrating corneal grafts. On the other hand, when one tries to insert corneal sutures into a corneal wound that is plugged by iris, the iris both obstructs the free passage of the needle and impedes the seizing of its tip and may also be damaged by the thread when it is pulled through, the fine fibres of the thread engaging the iris and twisting it, even when the silk seems ideally smooth. It is therefore preferable to disengage the iris from the wound first and to insert the sutures afterwards.

Summary

The results of repairing traumatic iris prolapses by replacement of the prolapsed iris have proved that this mode of repair is clinically sound and is to be preferred in suitable cases to the traditional method of abscission. A special technique described and recommended as a standard procedure consists essentially in an intra-ocular reposition carried out with a spatula through an incision at the limbus.

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DOMINANTLY INHERITED OPTIC ATROPHY*

BY

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LEBER'S disease, with its sudden onset in adolescence or early adult life, its rapid course, and puzzling mode of inheritance, has overshadowed other forms of inherited optic atrophy. The early literature contained many cases of "atypical" Leber's disease, but the continued emphasis on the established form, even in the monumental review by Bell (1931), hampered the emergence of other forms as separate clinical entities.

Waardenburg (1913) drew attention to the incidence of consanguinity in reported pedigrees of infantile optic atrophy suggestive of the existence of a recessive autosomal form. Rather more evidence for a dominant autosomal type is suggested by several reports in the early literature, such as the cases recorded by Knapp (1904), Lawson (1907), Nettleship (1909), Griscom (1921), Ginzburg (1923), Kawakami (1926), Alsberg (1927), Riedl (1935), and others; onset in childhood or infancy was common in such cases. Dominant optic atrophy is in fact not particularly uncommon; Lodberg and Lund (1950) have even suggested that in Denmark this type of optic atrophy is more common than Leber's disease. Recently attempts have been made to separate dominant optic atrophy into different forms. Thus Lodberg and Lund (1950) and Sorsby (1951) suggested a congenital and a post-natal form, while Jaeger (1954) suggested a third variety: dominant optic atrophy with nystagmus. The reports in the literature appear to fall into two main groups:

(1) The type which has been referred to as "congenital" has been infrequently reported. The largest family in this group, with twelve affected members, is that reported by Thompson and Cashell (1935) and by Dorrell (1932), which, as pointed out by Sorsby (1951), is the same family as that previously described by Nettleship (1909). Other similar families are those of Herzog (1930) and one of the three reported by Lodberg and Lund (1950). Although the affection has not in fact been seen at birth, Dorrell observed optic atrophy at 8 months. Some of Nettleship's patients speak of good vision up to the age of 3 or 4 years, although this in itself does not preclude the existence of optic atrophy as a congenital lesion. Visual acuity in these three families ranged from 6/24 to hand movements, the poorer levels of vision being common. Progressive deterioration appears to have been experienced by some patients, as in two cases recorded by Lodberg and Lund. Divergent strabismus and contraction of the peripheral field were noted in some members of each of these three families. Thompson and Cashell observed nystagmus in nine out of ten cases; a generalized pallor of the optic disc

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was a common finding in their cases, and was sometimes accompanied by attenuation of the retinal blood vessels. Taking these three family groups together—the Nettleship-Dorrell-Thompson-Cashell group, the Herzog family, and the Lodberg and Lund family—it is evident that, though there is a fairly definite pattern, there are considerable individual variations, especially in the degree of vision and the presence of nystagmus.

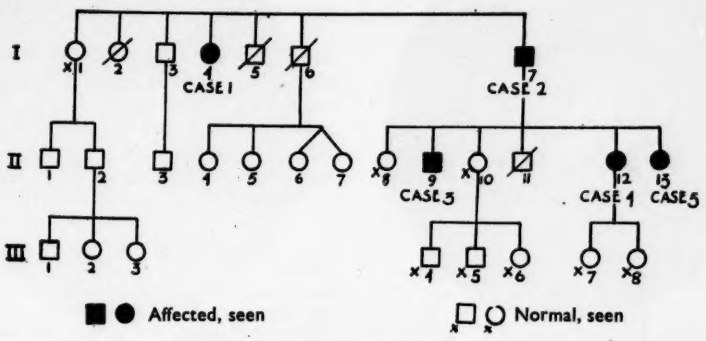
(2) The second type has been termed the "infantile" or "juvenile" form, or the "acquired" form. The largest study is that of Kjer (1956), who reported twenty families with at least 200 affected individuals. Kjer emphasized that there was wide variation in the degree of manifestation in any one family. Visual acuities ranged from 6/6 to 2/60, but vision was apparently never entirely lost. Progress with age is not constant. In 51 cases the disease appeared stationary, while in 43 there was slow progression. The age of onset is usually in childhood. The youngest case found by Kjer was aged 2 years, and by Scott (1941) 4 years. Lodberg and Lund (1950) give details of three siblings whom they re-examined while the disease was progressing. The first was passed as normal at the age of 7, but 3 years later there was marked temporal pallor and the visual acuity had fallen from 6/6 to 6/36. The second at the age of 9½ years showed temporal pallor and the visual acuity was recorded as 6/18; 4½ years later the visual acuity was reduced to 6/36 in the right eye and 6/24 in the left, with bilateral temporal pallor of the discs. The changes in the third were less striking; between the ages of 7 and 12 "light fading" of the temporal side of the disc became "pronounced" and the visual acuity fell from 6/24 to 6/36. Progressive deterioration with age is also suggested by the following analysis:

Visual Acuity in Better Eye	Scott (1941)		Kjer (1956)	
	Adults	Children	Adults	Children
6/6 –6/18	—	—	76	15
6/24–6/60	11	5	57	6
6/60–2/60	13	—	27	—
No data	—	—	7	12

Ophthalmoscopically, pallor is commonly confined to the temporal side, or is more marked on this side; in severe cases it is generalized. Contraction of the peripheral fields is rare, but in severe cases it may be gross, the temporal field being the more affected (Kjer). Enlargement of the blind spot was reported by Jaeger (1954), and a paracentral scotoma by Kjer. Poor colour vision and constriction of the colour fields was noted by a number of observers (Knapp, 1904; Scott, 1946; Lodberg and Lund, 1950; Jaeger, 1954). The reduction of the field for blue observed by Scott (1941) was confirmed by both Jaeger and Kjer, the latter drawing attention to the hemianopic nature of the colour fields, the temporal side being lost first. Kjer stressed the psychic anomalies and mentioned poor night vision.

PERSONAL OBSERVATIONS

(1) D. Family (Pedigree Chart I).



There were five affected persons in two generations:

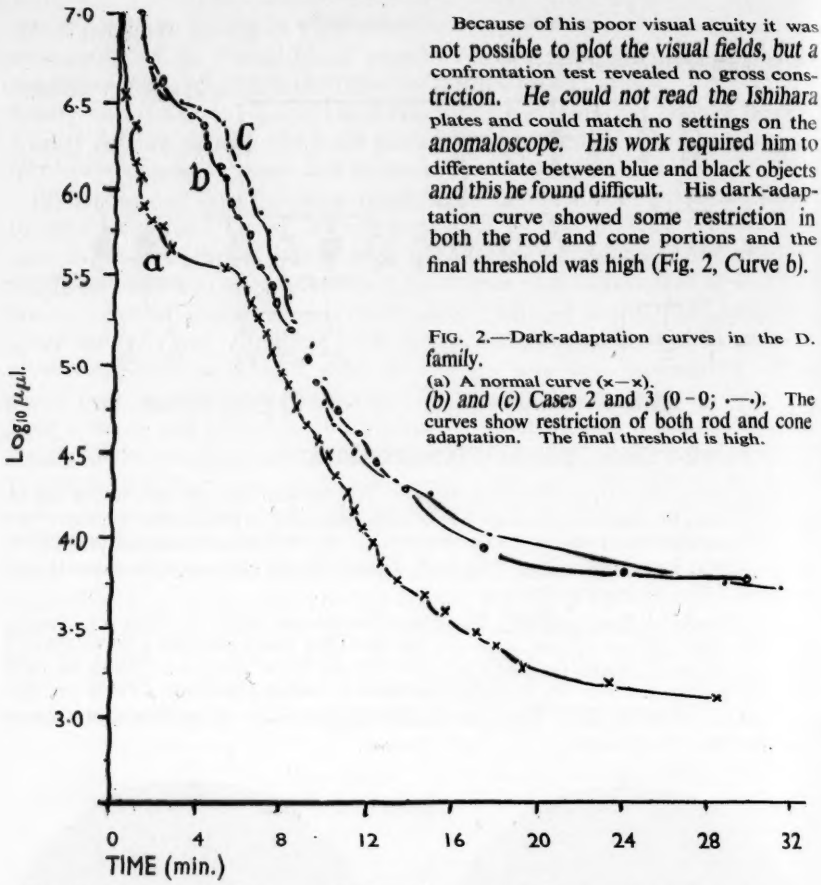
Case 1, Laura D—. (I, 4, Pedigree Chart I).—This patient was first seen at the age of 65 years when the visual acuity was 1/60 in each eye. She had had defective sight "all her life" and had always been "bad at colours". The Ishihara test was not practicable because of the low visual acuity. The fundi showed myopic changes at the maculae and temporal pallor of the optic discs.

Case 2, Frederick D—., aged 62 (I, 7).—The refraction was: Right +3 D sph., −2 D cyl., 95°; Left +4 D sph., −2 D cyl., 95°. He had had poor vision since his school-days and had been aware of a steady deterioration. In 1951 the visual acuity was 3/36 in the right eye, and 3/60 in the left. By 1956 this had fallen to counting fingers at 2 m. in the right eye, and at 1 m. in the left. The fundi showed marked pallor of the discs, especially on the temporal side (Fig. 1*a*).

(*a*) A severely affected man aged 62. The right disc had retained some pink colour on the nasal side, but otherwise the discs were grey. Visual acuity was reduced to counting fingers.

(*b*) A son (aged 37) and a daughter (aged 30) both had poor vision.

FIG. 1.—Appearances of the optic nerve in the D. family.



Because of his poor visual acuity it was not possible to plot the visual fields, but a confrontation test revealed no gross constriction. He could not read the Ishihara plates and could match no settings on the anomaloscope. His work required him to differentiate between blue and black objects and this he found difficult. His dark-adaptation curve showed some restriction in both the rod and cone portions and the final threshold was high (Fig. 2, Curve b).

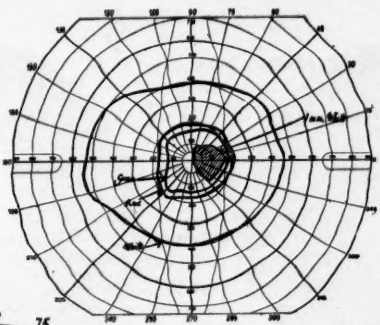
FIG. 2.—Dark-adaptation curves in the D. family.
(a) A normal curve (×—×).
(b) and (c) Cases 2 and 3 (0–0; ·—·). The curves show restriction of both rod and cone adaptation. The final threshold is high.

Case 3, Frederick John D—., aged 37 (II, 9), a son of Case 2.—His visual acuity was 6/36 in each eye in 1951 and 5 years later it had not altered. This man was illiterate and mentally subnormal, and as a child had been subject to epileptic fits. His response to the Ishihara test was poor. The anomaloscope showed him to have poor matching ability with a tendency towards protanopia. Colour fields were restricted, that for blue being absent. The field for white is shown (Fig. 3*a*, opposite), and the enlarged blind spot will be noted. His dark-adaptation curve was substantially the same as his father's (Fig. 2, Curve c). Temporal pallor was present in both optic discs (Fig. 1*b*).

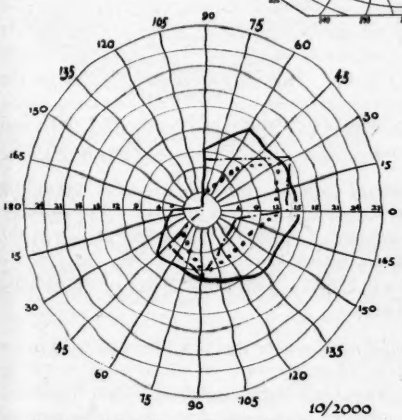
Case 4, Mrs. Christina G—., aged 30 (II, 12), a daughter of Case 2.—She had had poor vision since her school-days and was not aware that there had been any recent deterioration. For the past 5 years her visual acuity had been 6/60 in each eye. She failed completely on the Ishihara test and on the anomaloscope could only detect changes in luminance. As with her father (Case 2), it was not possible to plot fields of vision on a screen. Her dark-adaptation curve was normal except for a minimal restriction in the cone curve. Temporal pallor was present in the optic discs, but the nasal sides appeared normal (Fig. 1(*c*), opposite).

FIG. 1(*c*).—A son (aged 37) and a daughter (aged 30) both had poor vision.

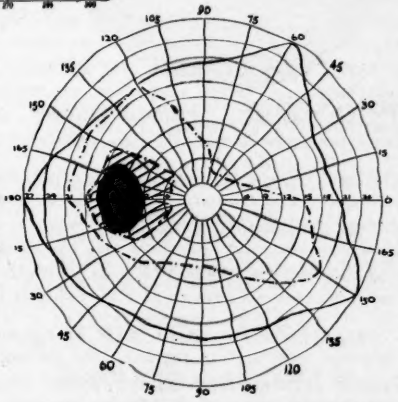
FIG. 1(*d*).—A younger daughter (aged 22) retained visual acuity of 6/24 and 6/18. Temporal pallor was present in all three siblings, but the coloration of the nasal side remained good.



(*a*) Case 3, right eye. The field for blue was absent, and the other colour fields were restricted. Note the enlarged blind spot.



(*b*) Case 5, left eye. Colour fields; note the severe restriction.

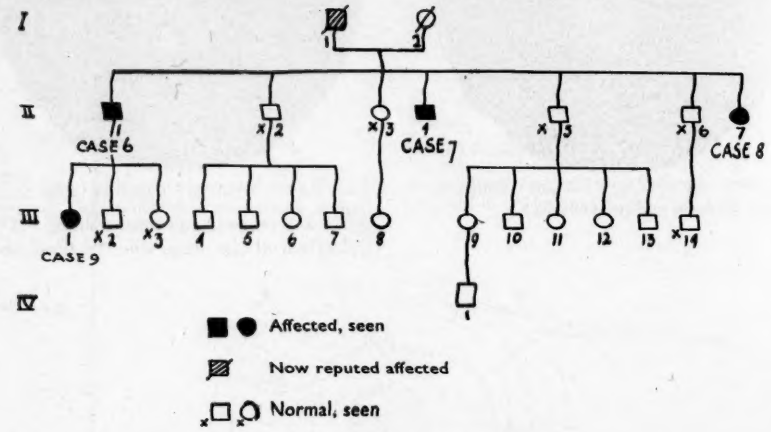


(*c*) Case 5, left eye. Fields for white; note the enlarged blind spot and restriction of the nasal field for a 2/2000 stimulus (—.—), while the 10/2000 field (—) was full.

FIG. 3.—*Fields of vision in the D. family.*

Case 5, Doreen D—., aged 22 (II, 13), proband.—The refraction was: Right −3·25 D sph., −0·5 D cyl., 65°; Left −3 D sph., −0·25 D cyl., 80°. In 1951 the visual acuity was 6/12 in each eye and this had fallen by 1956 to 6/24 in the right eye and 6/18 in the left. The colour vision was normal but the colour fields were severely restricted on the temporal side (Fig. 3*b*), the upper temporal quadrants of the fields being absent. The field for white showed an enlarged blind spot for a 2/2000 stimulus (Fig. 3c), and restriction of the nasal field. The optic discs showed a mild temporal pallor (Fig. 1*d*).

(2) P—. Family (Pedigree Chart II).



There were four affected persons in two generations:

Case 6, John Alfred P—., aged 57 (II, 1, Pedigree Chart II), proband (Royal Eye Hospital record number, 33/1104).—He had had poor sight since his school-days and in 1916 was rejected by the Army on this account. In 1933 his visual acuity was 6/60 in each eye and by 1946 this had fallen to 3/60 in the right eye and 4/60 in the left; 5 years later it was 3/60 in each eye. The fields for white had not altered markedly between 1933 and 1951 (Fig. 4, opposite). Colour vision was almost completely lacking in 1951 but it was possible to plot a small field with a blue stimulus (Fig. 4). This response was present on the nasal side only. Dark adaptation was markedly restricted both for rod and cone vision (Fig. 6*b*, overleaf). Temporal "atrophy" of the optic discs was observed in 1933, and this is now considerable (Fig. 5*a*, opposite) but some colour remains on the nasal sides.

Case 7, George P—., aged 45 (II, 4), brother of Case 6.—His visual acuity was 6/9 in each eye. The Ishihara test showed complete colour blindness and the anomaloscope showed the hue discrimination to be much reduced. Pallor of the temporal side of the discs was observed and there was the same considerable reduction in his night vision (Fig. 6*c*, overleaf) as was seen in his brother (Case 6). This patient was uncooperative and it was not possible to plot his fields of vision.

Case 8, Lilian P—., aged 34 (II, 7), sister of Case 6.—Her visual acuity was 6/24 in the right eye, and 6/18 in the left. At the age of 18 years she was admitted to a mental hospital suffering from schizophrenia. She knew her sight had always been poor and that she was colour blind. However, she thought that when she first entered the hospital she could see colours better. She failed completely on the Ishihara test. It was not possible to plot her visual fields but the manner in which she turned her head to the side

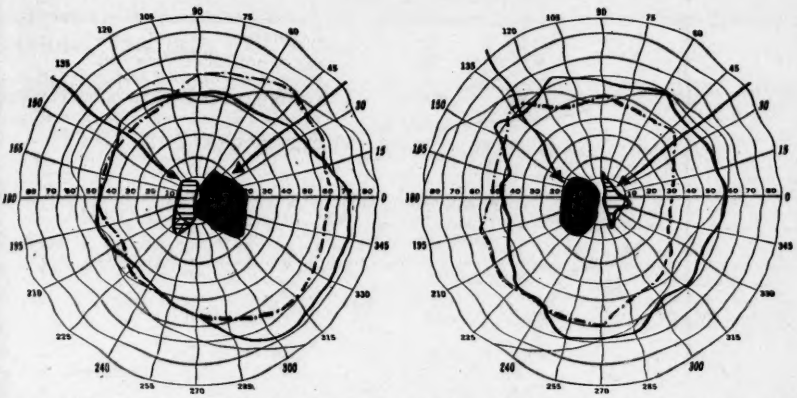


FIG. 4.—*Case 6, right and left eyes. The colour fields show that only a small field for blue* remained on the nasal side. The fields for white show enlargement of the blind spot. The peripheral fields were first plotted in 1933 (—) and, as will be seen, were little changed on examination in 1951 (—·—).

FIG. 5 (*a*).—*Case* 6, a man aged 57 with 3/60 vision each eye, left eye. Some pink coloration was still present on the nasal side of the disc.

FIG. 5 (*b*).—*Case* 8, a sister, aged 34, of Case 6, left eye. The nasal side of the disc still retained some pink coloration. Visual acuity 6/18.

when she wanted to regard an object was suggestive of a central scotoma. The optic discs showed considerable temporal pallor (Fig. 5*b*).

Case 9, Rose P—., aged 26 (III, 1), daughter of Case 6.—She attended the Royal Eye Hospital in 1946 (46/3713), when the visual acuity was 6/18 in each eye. The visual fields were normal and the discs were pale, but not so pale as her father's. Defective vision had been first noticed at the age of 15 years.

Analysis

Mode of Inheritance.—In the D. family affected individuals occur in two generations. In the first generation, two out of the three individuals seen were affected. There had been seven in this sibship, but three are now dead and a fourth could not be traced. All the six children of I, 7, an affected

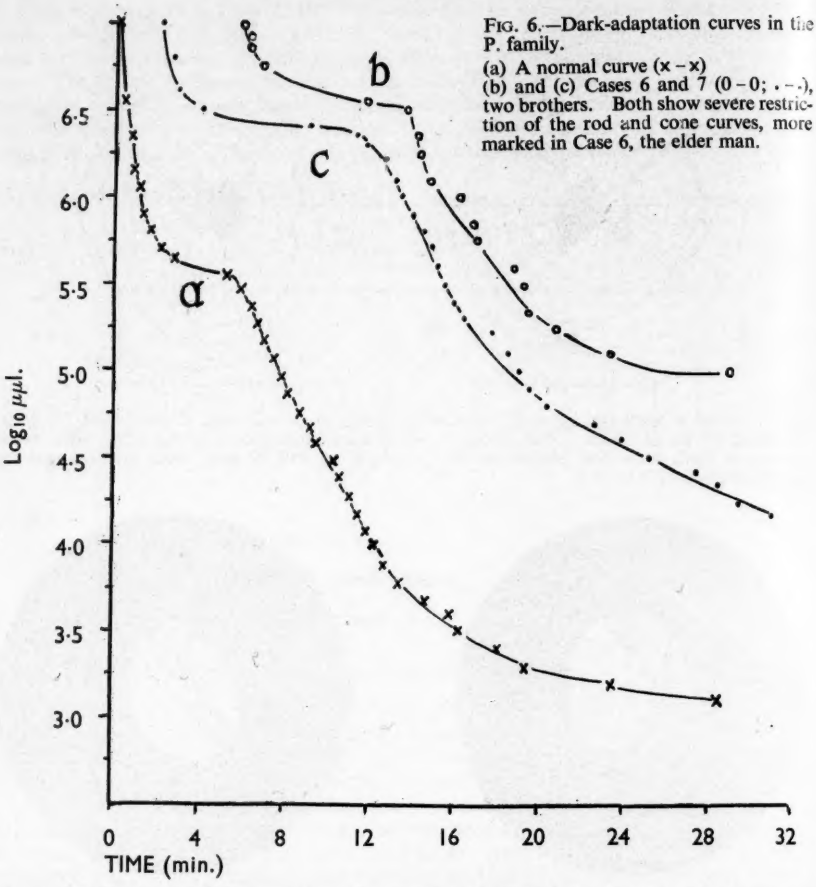


FIG. 6.—Dark-adaptation curves in the P. family.
(a) A normal curve (x – x)
(b) and (c) Cases 6 and 7 (0 – 0; . – .), two brothers. Both show severe restriction of the rod and cone curves, more marked in Case 6, the elder man.

man, were seen, with the exception of II, 11, who was killed while flying in the R.A.F. in 1944 aged 22; and whose sight was presumably normal. Of these six siblings, three were affected and three normal (if II, 11, be included as a normal). The two children of II, 12, an affected woman, aged 5 and 7 years, are both normal.

In the P. family two generations included four affected individuals. There was hearsay evidence which suggested that I, 1, had also been affected (his sons reported that he had had poor sight all his life and was unable to learn to drive on account of this defect). Generation II included seven individuals of whom three were affected. In Generation III one woman was affected; she was one of three children of an affected man.

The pattern of inheritance seen in both families strongly suggests dominant autosomal inheritance. If I, 1, in Pedigree Chart II is accepted as an affected

individual, then the evidence for dominance is conclusive. Sex-linkage is obviously excluded.

Visual Acuity.—The nine cases are arranged according to age in the Table.

TABLE
VISUAL ACUITY IN RELATION TO AGE

Case No.	Examination								
	First			Second			Third		
	Age (yrs)	Visual Acuity		Age (yrs)	Visual Acuity		Age (yrs)	Visual Acuity	
		Right	Left		Right	Left		Right	Left
5	17	6/12	6/12	22	6/24	6/18			
4	25	6/60	6/60	30	6/60	6/60			
9	26	6/18	6/18						
3	32	6/36	6/36	37	6/36	6/36			
8	34	6/24	6/18						
6	39	6/60	6/60	52	3/60	4/60	57	3/60	3/60
7	45	6/9	6/9						
2	57	3/36	3/60	62	Finger counting at 2 m.	Finger counting at 1 m.			
1	65	1/60	1/60						

(*a*) The visual acuity in individual cases tends to diminish although at varying rates. In 5 years Cases 2 and 5 showed an appreciable reduction while Cases 3 and 4 showed no change. Case 6 showed a gradual reduction over 18 years.

(*b*) The visual acuity of different cases bears little relation to their age. Case 7 had visual acuity of 6/9 at the age of 45, while Case 4 had visual acuity of 6/60 at the age of 25.

Fundus Appearances.—Pallor of the optic disc was the only ophthalmoscopic sign observed. This ranged from temporal pallor with good coloration on the nasal side, as in Cases 4 and 5, to a more general whitening of the discs as in Cases 2 and 6. The amount of pallor is only an approximate guide to the degree of visual handicap.

Dark Adaptation.—Poor night vision was found in five affected individuals, in one of whom the defect was minimal (Case 4). The dark-adaptation curves for the remaining four are given in Figs 2 and 6. The two cases in the P. family showed a marked restriction of both cone and rod vision (Fig. 6, Curves b and c). The point at which rod vision became more

sensitive than cone vision was delayed by some 5 minutes. Case 7 had good visual acuity (6/9), but the other cases with poor night vision had low visual acuity.

Colour Vision and Colour Fields.—Of the eight cases examined by the Ishihara and anomaloscope tests, seven showed evidence of defective colour vision. The one case with normal colour vision, a girl aged 22, showed a quadrantic, almost hemianopic field defect. It was possible to plot the colour fields in only two other cases and these were severely constricted. In Case 3 only the plotting of red and green fields was possible, the blue field being absent (Fig. 3*a*), and in Case 6 only the blue field could be plotted (Fig. 4). This last was an unexpected finding, since other workers have emphasized early loss of the blue field with the retention of the red and green fields. The hemianopic shape agrees with previous findings. In three patients low acuity and gross colour vision defects rendered the procedure impracticable. Of the rest, two declined to be tested and another was a patient in a mental hospital which lacked the necessary equipment.

Fields for White Stimuli.—In the three cases in which fields for white were plotted an enlargement of the blind spot was observed (Figs 3*a*, 3*c*, and 4). The periphery of the field is not markedly contracted. In Case 6 the fields for white were plotted in 1933 and again in 1951 and the change was small; in the same period his visual acuity fell from 6/60 to 3/60 in each eye. In Case 2 a confrontation test revealed no marked constriction of the periphery of the fields, although the visual acuity at the time was only 3/36 in the right eye and 3/60 in the left.

Psychic Abnormalities.—These were noted in one patient in each family, and one of these was a patient in a mental hospital.

Discussion

(1) Onset and Course.—Like previous studies, the present records show dominant optic atrophy to be an insidious affection. In two of our cases the anomaly was not discovered until the ages of 15 and 17, when the visual acuity was 6/18 and 6/12 respectively. The success with which patients overcome their visual difficulties is possibly accounted for by the fact that, although their visual acuity may be low, the periphery of their fields is largely intact. In addition, the gradual reduction in visual acuity, unaccompanied by any marked contraction in the field for white allows the patient to become adapted to his failing vision. Even though his vision was below 6/60, Case 2 was still working. In three of the six cases examined on more than one occasion a fall in visual acuity was detected. Since one of these is 22 years of age and another 62 it seems probable that a gradual diminution occurs throughout life.

(2) **Functional Disturbances.**—Defective colour vision has been reported by several other observers as one of the more consistent findings among affected adults and this was the case in at least seven of our patients. The contraction of colour-fields stressed by Kjer (1956) as especially marked on the temporal side and most definite for blue was also found in two of those patients who could be examined. In a third (Case 6) an anomalous finding was the persistence of a somewhat hemianopic field for blue when all other colour fields had already been lost.

Kjer noted defective night vision in some of his cases. The present study confirms that this defect is not an uncommon feature. The anomaly is more than a mere raising of the final threshold. The curves of both cone and of rod adaptation are late in appearing and both are restricted.

(3) **Psychic Abnormalities.**—These were noted by Kjer in about 10 per cent. of his cases, and were seen in two cases in the present study.

(4) **Outstanding Issues.**—The significance of poor dark adaptation in dominant optic atrophy still requires elucidation. Whether involvement of both rod and cone curves is characteristic of this particular affection or an aspect of any optic atrophy needs to be determined.

The present case reports show how difficult it is to establish in any particular family whether the affection is congenital or post-natal in origin, and whether indeed this distinction with its implications as to severity and prognosis is valid. Investigations are needed in suitable families specially studied for this possible distinction.

Summary

Two families with inherited optic atrophy were examined. The following characteristics were observed:

(1) The mode of inheritance is almost certainly dominant and autosomal.

(2) The visual acuity of affected adults diminishes slowly throughout life.

(3) The visual fields show an enlarged blind spot, a good peripheral field for white, but contracted fields for colour, usually especially marked for blue (an exception was a case in which the blue field was retained while those for red and green were lost). The field defect may be quadrantic or hemianopic, the temporal side being the more severely affected.

(4) Colour vision among the adults is usually defective or absent.

(5) Poor dark adaptation is often seen, usually with a marked restriction of both the rod and the cone portions of the dark-adaptation curve.

(6) Pallor of the disc, most marked on the temporal side, is a constant finding in affected individuals, but the degree of pallor is only an approximate guide to the amount of visual defect.

We are indebted to Professor Arnold Sorsby for his encouragement in the preparation of this paper and to Mr. D. W. A. Mitchell of the London Refraction Hospital for referring to us the first of the two families reported.

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CASE NOTES

INCOMITANT STRABISMUS OF UNUSUAL ORIGIN*

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Case Reports

Case 1, a 16-year-old Hindu girl, was admitted to hospital on March 14, 1955, complaining of diplopia and turning in of the left eye for the last 10 months. The symptoms followed a wasp sting on the lid below the inner canthus (by the common wasp *Polistes habreus*). Acute, intense oedema of the lids and conjunctiva lasted for 3 days, during which the eye was completely closed. When the swelling subsided and lids could be opened, the patient experienced diplopia, particularly on looking to the left, and the left eye was turned in. Injections of vitamin B_1, cod liver oil by mouth, and an occlusive bandage over the right eye were ordered, but she gave up treatment after 30 days, and the symptoms persisted.

There was nothing relevant in the past history. Symptoms of asthenopia had been relieved by a −2D sph. correction 5 years previously. Nothing significant was noted in the family or personal history.

Examination.—The left eye was convergent 25°, as tested with the pen-torch. There was slight limitation of laevo-version with increase in diplopia on looking to the left.

The visual acuity in each eye was 6/36 J.1, and 6/9 J.1 with −2 D sph.

Synoptophore

Major Amblyoscope	15° to left	Ahead	15° to right
Fixing right	26°	25°	24°
Fixing left	30°	27°	26°

The Hess screen test showed limited action of the left external rectus muscle.

Diagnosis.—Left external rectus paresis.

Operation.—Advancement of the left external rectus muscle, with 5 mm. recession of the left internal rectus muscle.

Result.—On the 15th day after operation the eyes were parallel, but there was slight diplopia on extreme laevo-version

Synoptophore

Major Amblyoscope	15° to left	Ahead	15° to right
Fixing right	3°	1°	1°
Fixing left	5°	2°	2°

The range of fusion at this time was only 4°. After fusion exercises for about 6 weeks this increased to 25°.

The patient has not reported back for further examination.

* Received for publication May 13, 1957.

Comment

The case was clearly one of left external rectus paresis following a sting. The formic acid thus injected results in an acute allergic and inflammatory reaction in the tissues (probably due to liberation of histamine) leading to accumulation of a toxic transudate in the affected areas. The transudate, in this case, diffused into the conjunctival sac, and the conjunctival tissues shared the inflammatory reaction. The abducens nerve endings were affected by the toxic contents of this transudate, and this led to paresis of the external rectus muscle. This nerve has a peculiar predilection for giving up its function on the slightest pretext. Probably this is the weakest nerve and cannot stand as much strain as the others, which would account for the solitary involvement of the external rectus in this case. The deficient range of fusion may have been a contributory factor.

Case 2, a 25-year-old man, a record keeper in a local office, had had aches and pains in the outer part of left eye for the last 6 months. The symptoms increased on dextro-version, and the effort produced diplopia which increased with the increased attempt to move the eye in this direction. About a year ago there had been an episode of acute conjunctivitis, with acute redness and swelling of the conjunctival sac and lids for 4 to 5 days. Though antibiotic therapy gave appreciable relief, a patch of congestion remained at the outer canthus under the outer part of the upper lid. A nodular swelling had started to develop at this site and in the last 4 months had assumed the size of a large pea, and an outward deviation of the left eye was noted. The patient had eaten pork about 10 years before and again one year ago.

Examination.—There was a 7° divergent strabismus in the left eye. Beneath the outer third of the upper lid, above the outer canthus, was a tense, pinkish-grey, sub-conjunctival swelling the size of a pea. It was tender to the touch, and around it the conjunctiva was congested for an area of about 4 mm. (Fig. 1).

The other ocular structures and the fundus were normal. The visual acuity was 6/6 J.1 in each eye and there was no error of refraction. The eye movements were normal in all directions, except for the difficult and painful extreme dextro-version.

FIG. 1.—Appearance of Case 2 before operation.

Synoptophore

Major Amblyoscope	15° to left	Ahead	15° to right
Fixing right	L/R 1Δ	L/R 1Δ	L/R 1Δ
Fixing left	L/R 1Δ	L/R 1Δ	L/R 1Δ
	−2°	−3°	−7°

The Hess screen revealed limitation of action of the left internal rectus and over-action of the right external rectus.

The total leucocyte count was 8,700 per cent., polymorphs 69 per cent., lymphocytes 24 per cent., monocytes 1 per cent., eosinophils 6 per cent.

Operation.—The cyst was removed under local anaesthesia. It was found embedded

in Tenon's capsule, lying in contact with the upper border of the left external rectus muscle. It was ovoid, pearly-white in colour, about 4 mm. across, with a dense white spot near the narrow end resembling a typical cysticercus cellulosæ; a probable diagnosis was made on the operation table.

Histological Report (by Dr. Gurbachan Singh of the Pathology Department).—Microscopic examination of the cyst showed a typical characteristic appearance of a double-walled connective tissue capsule. The outer wall of the capsule showed infiltration with chronic inflammatory cells. Its inner surface was lined with the continuation of the epithelium of the tortuous canal of the body of the larva (Fig. 2).

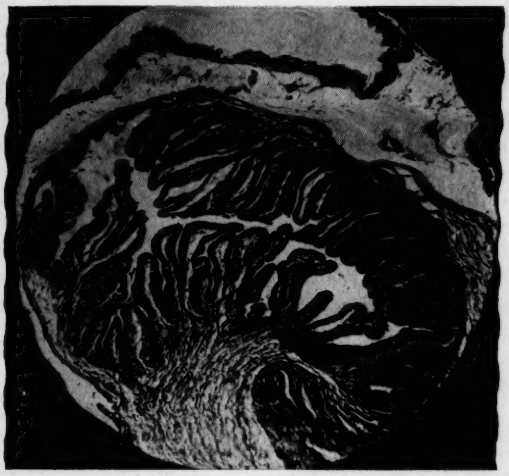

FIG. 2.—Histological appearance of cyst in Case 2, showing body of larva. In this section, the head and the neck of the larva were not discernible.

Radiological examination of the other parts of the body revealed no calcified cysts.

Result.—There was marked relief of the diplopia on the seventh post-operative day, and on the 15th day the following measurements were made:

Synoptophore

Major Amblyoscope	15° to left	Ahead	15° to right
Fixing right	L/R 2½Δ	L/R 2½Δ	L/R 2½Δ
Fixing left	L/R 2½Δ −1°	L/R 2½Δ −1°	L/R 2½Δ −1°

3 months after the operation the eyes were parallel in all directions of gaze, with unrestricted movement in all directions. There was no diplopia and a normal Hess chart.

Synoptophore

Major Amblyoscope	15° to left	Ahead	15° to right
Fixing right	L/R 2½Δ	L/R 2½Δ	L/R 2½Δ
Fixing left	L/R 2½Δ	L/R 2½Δ	L/R 2½Δ

Comment

Ocular cysticercus cellulosae is a rare condition. Duke-Elder (1952) refers to 372 cases of ophthalmic infections by such cysts in the ocular structures.

The lesion may appear as a localized hemispherical swelling anywhere in the ocular structures, commonly at the lower inner angle of the eye in the

lower fornix, and is sometimes intra-ocular. There is a severe inflammatory reaction with chemosis of the conjunctiva, oedema, and much pain. Ptosis, optic neuritis, and paresis of external muscles may also occur.

In the case reported above the symptoms came on one year after eating pork. The initial severe inflammatory reaction subsided leaving behind a congested patch at the outer canthus. At operation the cyst was found embedded in Tenon's capsule and adherent to the sheath of the external rectus muscle. This muscle was consequently in a state of irritation, and this resulted in constant overaction and consequent paresis of the internal rectus leading to a divergent strabismus of 7°. This theory of the cause of the strabismus is proved by the relief of the strabismus and diplopia which resulted from the removal of the cyst.

The constant hyperphoria of 2△ may probably have been a pre-existing one. This was probably reduced by the effect of the cyst weighing down on the eye.

Summary

(1) Two cases of incomitant strabismus are reported.

(2) In one case, the cause was a toxic neuritis of the sixth nerve caused by toxins in the inflammatory transudate produced from a sting of the common wasp. Surgery was followed by relief of both diplopia and strabismus.

(3) In the second case, the strabismus was due to an irritative spasm of the external rectus muscle caused by a cysticercus cellulosae which was embedded in Tenon's capsule in relation to the sheath of the muscle. Removal of the cyst was followed by a relief of both strabismus and diplopia, the main subjective symptoms in the case. A pre-existing hyperphoria had been diminished by the weight of the cyst.

We are grateful to Dr. G. S. Malhotra, for giving fusion exercises to Case 1.

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INFECTION WITH MICROCOCCUS TETRAGENUS AS A COMPLICATION OF PENETRATING KERATOPLASTY*

BY

G. J. ROMANES

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EXPERIENCE in a recent case shows many points of interest.

Case Report

A male medical student, aged 22, had bilateral keratoconus which had been successfully treated by Mr. H. Treissman with contact lenses for 7 years. Recently the left eye had become intolerant to the contact glass with recurrent corneal ulceration and scar formation. The patient was referred to Mr. B. W. Rycroft for consideration of keratoplasty.

History.—There were no points of relevant ophthalmic interest in the general health or family history. There was no evidence of closed focal sepsis and a routine radiological examination of the chest was normal.

Examination.—Visual acuity: right eye with contact lens 6/12 (ii); left eye with contact lens 1/60.

Projection of light was accurate and brisk in each eye to a dull stimulus. The intra-ocular pressure was normal. Marked cone formation was present on each cornea with typical ring distortion and concentric concentration was seen on the keratoscope. There was fluorescein stain on the left cornea limited to the apex of the cone about 3 mm. in diameter; it was surrounded by a circle of punctate staining (Fig. 1).

FIG. 1.—Left eye on admission.

Slit-Lamp Report (Left eye).—The epithelium was regular at the periphery, but in the central region there was marked irregularity with some opacity. The contour of the anterior surface was obviously conical with the apex slightly down and in from the corneal centre.

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The stroma had normal thickness above and below at the periphery, but there was a horizontal thin area across the cornea. In the cone the total stromal thickness was reduced by 70 per cent. compared to the periphery above and below.

The posterior corneal surface showed marked irregularity with linear wrinkling of Descemet's membrane in the base of the cone. There were no cells adherent to the posterior corneal surface.

The anterior chamber was increased in depth in the centre, and was normal at the periphery; no flare was present. The iris pattern and movements were normal. The lens capsule and anterior lens cortex were normal. The fundus showed no abnormality with the ophthalmoscope.

Plan.—Because of recurrent ulceration, diminished contact lens tolerance, deterioration of visual acuity, and the progressive nature of the disease, a left penetrating keratoplasty was advised.

Donor Material.—This was a fresh eye, excised with full aseptic precautions 2 hours previously for a choroidal neoplasm, and suspended in saline vapour in the refrigerator at +4° C. This eye had been treated for 48 hours before operation with chloramphenicol 1 per cent. ointment. An overnight pad on the donor eye showed no stickiness and a control 48-hour culture of the conjunctiva was sterile.

Operation.—On August 28, 1956, an 8-mm. penetrating keratoplasty (B. W. R. and G. J. R.) was performed under local anaesthesia; intravenous potentialization was obtained with Pethidine, Phenergan, and Largactil (dosage 50 mg. of each).

The graft was secured by four direct sutures in the vertical and horizontal meridians, reinforced with two oblique overlay sutures across sterile egg membrane. At the end of the operation a subconjunctival injection of 10 mg. cortisone was given. Ung. chloramphenicol 1 per cent. and ung. eserine 0·5 per cent. were applied after the anterior chamber had reformed. Air was not insufflated. Double pad dressings were applied with a pressure bandage over the left eye.

Post-operative Course.—There was a little vomiting in the first part of the week but there was no undue discomfort in the eyes. The first dressing was done on the seventh day, as is the routine custom. The pad was sticky with a yellowish-green purulent material and there was excessive swelling of the lids. The lower lid Frost suture was under tension due to the oedema of the lids. After local anaesthesia, the eye was seen to be red with gross conjunctival infection and chemosis; one cruciate suture had cut out in the infra-nasal region. The cruciate sutures and egg membrane were removed and pus was adherent to the direct sutures; the graft showed infection of the infra-nasal quadrant extending almost to its centre, and the 6 and 9 o'clock direct sutures had also cut out. There was also gross infection of the host stroma with a 2-mm. hypopyon. The pupil was in miosis with many posterior synechiae.

A swab was taken, an immediate subconjunctival injection of streptomycin and penicillin was made, and "Polyfax" ointment was instilled. Local intensive antibiotic therapy was immediately commenced with a drop of streptomycin 50 mg./ml. every minute.

Culture Report.—A pure growth of *Micrococcus tetragenus* sensitive only to chloramphenicol was found.

> "*Micrococcus tetragenus* may occur as a commensal on the mucous membrane of the upper respiratory passages; it has also been found on the skin. It is often found in suppurations in the region of the mouth or in the neck, *e.g.* dental abscess, and also occurs in various lesions of the respiratory tract, in phthisical cavities, abscesses in the lungs, etc. Sometimes it is present alone, and probably has a pyogenic action in the human subject under certain conditions. In most cases it is associated with other organisms. Cases of general infection along with pneumonic symptoms have been recorded, the organism having been isolated from the blood; recovery was the rule. Cases of pyaemia have also been described in which this

organism was found in a state of purity in the pus in various situations." (Browning and Mackie, 1949.)

An examination under general anaesthesia was made the next day, and it was found that the union of the host-graft junction was weak and primary ectasia of the graft had set in. During a subconjunctival injection of saturated solution of penicillin and streptomycin, the wound gave way below with loss of the anterior chamber. To reinforce this rupture a conjunctival "bucket-handle" flap was immediately fashioned and secured over the bulging section to provide cover and additional blood supply. When the flap was removed a week later the union was seen to be firm again.

On the 8th day there was a small leucocytosis. Daily subconjunctival chloramphenicol and mydricaine injections were continued for 8 days under light general anaesthesia. During the whole of this period chloramphenicol 250 mg. daily was given by mouth, and injections of streptomycin 1 g. daily and penicillin 1 M units were given daily. Chloramphenicol drops were instilled at 5-minute intervals day and night. Phenylephrine 10 per cent. was used for the first 2 days; thereafter it was discontinued and gutt. cortisone 5 mg./ml. were started. Atropine 1 per cent. drops were used every 4 hrs throughout.

On the tenth day the total white cell count had fallen to normal, indicating that the infection had now responded to the antibiotics.

The clinical appearance showed progressive changes of improvement from a state of gross infection with hypopyon to a final stage of a white eye with a small colarette anterior synechiae in the inferior nasal quadrant. There remained little more than peripheral fibrosis in the graft; the central area was clear and there was minimal residual opacity in the adjacent host tissue.

The visual acuity in the left eye was counting fingers at 1 m. 4 weeks after operation and in the seventh week it was 6/36 unaided, and the patient was discharged from hospital.

Post-operative Visual Acuity.—6 weeks later the visual acuity in the left eye without correction was 6/18, and the eye was white and quiet. A contact lens trial 32 weeks after operation resulted in a visual acuity of 6/9 and the tolerance was excellent (Fig. 2).

FIG. 2.—Left eye 32 weeks after operation.

Comment

This case represents one of only two infected grafts in over 300 graft cases at this Unit. The following points are of interest:

(1) A fresh donor eye, in spite of previous intensive treatment by antibiotics,

may be a potential source of infection owing to chance resistance. It is not possible, however, to define the exact source of infection in this case, but the initial site appeared to be in the graft. The organism rarely causes pyaemic lesions and is usually considered to be a commensal in an established respiratory lesion. The theatre is air-conditioned and has regular bacteriological control.

(2) Early bacteriological diagnosis and establishment of sensitivity enabled appropriate antibiotic therapy to be given at the most opportune time.

(3) The opinion of our pathologist, Dr. A. Sachs, that little fibrotic reaction would result from this particular organism has been fully justified. The visual acuity improved from 1/60 to 6/9.

(4) Intensive and continuous local treatment, coupled with devoted and enlightened nursing, were the factors of prime importance in the successful outcome. "Minute drops" were used for 24 hrs, and one drop every 5 minutes for 3 days and nights.

(5) The prognosis of severe intra-ocular post-operative infection need not necessarily be catastrophic to vision, provided skilled team-work is available.

I wish to acknowledge the help and encouragement I have had in the preparation of this report from Mr. B. W. Rycroft and Dr. A. Sachs, and to place on record the devoted care of Sisters King and Nuthall and the nursing staff of the Corneo-Plastic Unit. I am indebted to Gordon Clementson for the photographs.

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'AMYLOID TUMOUR' OF THE EYELIDS*

BY

S. AGARWAL AND J. B. SHRIVASTAV

From the Department of Pathology and Bacteriology, Medical College, Nagpur, India

AMYLOIDOSIS is usually categorized as a degenerative process. Nevertheless, the nature and pathogenesis of this disorder remain most uncertain. Reimann, Koucky, and Eklund (1935) have classified amyloidosis in four groups: primary, secondary, tumour-forming, and associated with multiple myeloma.

Tumour-forming amyloidosis is known to occur as a primary process in the tissues of the eye, urinary bladder, tongue, and upper respiratory passages. Instances of localized amyloidosis involving ocular tissues after chronic inflammation are rare, and only a few have been recorded in the recent literature (Elles, 1945; Chinaglia, 1952; Handousa, 1954; Oppel, 1956).

A review of the literature suggests that amyloidosis as such is uncommon in India. Mathur and Bhende (1957) could trace only eighteen reported cases and added two cases of their own. In none of these cases was involvement of the ocular tissues described and to our knowledge no case of tumour-forming amyloidosis has been reported from India, although chronic inflammatory conditions of the eye are common.

Case Report

A woman aged 30 years complained of a gradually increasing swelling of the upper and lower lids of both eyes for the last 12 months and of difficulty in seeing. The swelling of the lids started from the inner angles and had involved the right side more than the left.

Examination.—The right side showed a uniform thickening of the upper and lower lids with a well-defined firm round nodule nearly 0·5 cm. in diameter on the medial third of the upper lid. The left eye also showed uniform thickening of both the lids and a similar nodule occupying the medial third of the upper lid (Fig. 1).

FIG. 1.—Bilateral amyloid tumour of the eyelids.

The nodules were not tender and were not adherent to the skin. A detailed examination of the eyes was not possible owing to the narrowed palpebral fissures. The palpebral conjunctiva could not be examined as it was not possible to evert the lids. Absence of

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epiphora suggested that the naso-lacrimal ducts were not involved. The patient denied having applied or injected any irritant to the eyes, and there was no history of any chronic infection in the eye or elsewhere.

General examination revealed no abnormality. The blood picture and urine were normal and serological tests for syphilis were negative.

A clinical diagnosis of "trachomatous folliculoma" or "fibromatous tumour" was made, the nodules were excised, and the lids were corrected.

Pathology.—The tissues consisted of four irregular pieces, each measuring nearly 0·5 cm. in largest dimension. They were uniformly yellowish in colour and of firm consistency. There was no obvious capsule around them. One of the cut pieces was treated with iodine which stained it brown and the colour changed to blue on addition of sulphuric acid thus suggesting the presence of amyloid in the tissue.

Histologically, the nodules were composed of large masses of a homogeneous, non-fibrillar, eosinophilic hyaline material lying beneath the conjunctival epithelium (Fig. 2).

FIG. 2.—Masses of amyloid beneath the conjunctival epithelium, with foci of chronic inflammatory cells. Haematoxylin and eosin × 100.

The deposition was diffuse but predominated around the blood vessels (Fig. 3, opposite).

The lumen of the blood vessels was markedly narrowed and often occluded because of the infiltration of the hyaline material into the vessel walls. The acini of the glands of Moll were separated by the infiltration of similar substance into the interstitial tissue around them (Fig. 3).

The adipose tissue of the lids also showed the hyaline substance around the walls of the vacuolated fat cells (Fig. 4, opposite). The epithelium of the palpebral conjunctiva showed thickening and squamous metaplasia in some places while in others it was thinned out. The epithelium also showed gland-like invaginations. Chronic inflammatory cells in which lymphocytes and plasma cells were prominent were seen lying in groups below the conjunctival epithelium, round about the blood vessels and within the diffuse hyaline mass of amyloid tissue. There was no cholesterol deposition or calcification, and no giant cell reaction. The hyaline masses showed the typical metachromatic reaction with Congo red which stained it pinkish-red.

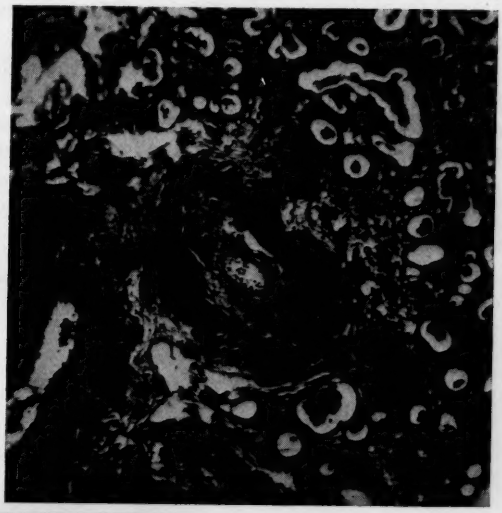

FIG. 3.—Deposition of amyloid around blood vessels and in interstitial tissue around the glands of Moll. Haematoxylin and eosin ×100.

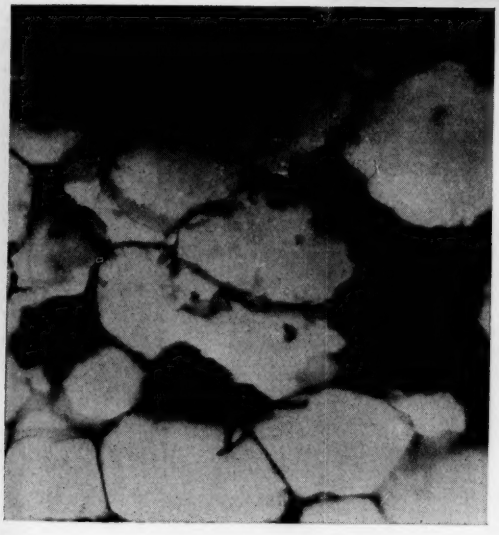

FIG. 4.—Amyloid around fat cells. Haematoxylin and eosin × 300.

Comment

The histological appearance and specific tinctorial reactions for amyloid show that the nodules were of localized amyloidosis of the eyelids.

The aetiology of amyloid degeneration of the ocular tissues is obscure. According to Oppel (1956), these tumour-like degenerative conditions develop on the basis of chronic conjunctivitis, and trachoma is present in 60–70 per cent. of such cases. In the cases of amyloidosis described by other

authors inflammatory conditions of the eye also preceded the degenerative change.

Lubarsch (1929) first clearly defined the difference between primary and secondary amyloidosis, the following being characteristic of primary amyloidosis: absence of antecedent or co-existent disease, involvement of mesodermal tissues (such as smooth and skeletal muscles, cardiovascular system and skin rather than liver, spleen, kidneys and adrenals), variability in the staining reactions of amyloid as opposed to their constancy in the secondary form, and a tendency to nodular deposition of amyloid.

It has been noted that primary and secondary amyloidosis are not sharply defined terms and considerable overlapping of the characteristic features described for each of them occurs in some cases (Koletsky and Stecher, 1939; King, 1948).

In the present case there seems to be an overlapping of these features. The localized nature of the tumour and the involvement of the mesenchymal tissues is in favour of primary amyloidosis, while the well-marked specific staining reaction for amyloid, association of chronic inflammatory cells, thickening of the lids, and absence of giant cell reaction around the masses of amyloid (Iverson and Morrison, 1948) point towards secondary amyloidosis. The primary inflammatory process in this case was probably trachoma—a condition frequently seen in India.

We are grateful to Mr. S. P. Sookraj for supplying the photograph of the patient.

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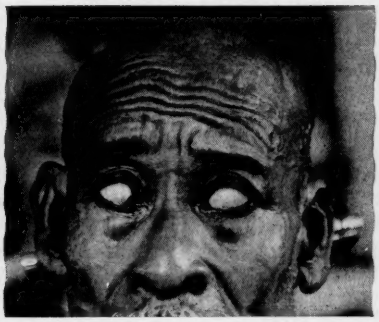
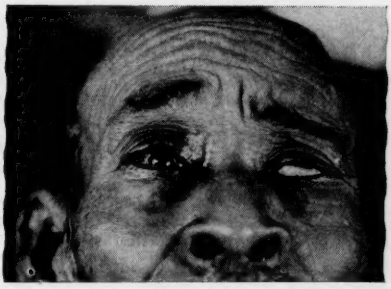
TWO CASES FROM MALAYA*

The following case notes, received from Dr. B. M. McOwan, of the General Hospital, Penang, Malaya, are unusually brief, but are being published for the sake of the drama of the illustrations.

(*1*) *Strabismus Fixus.*—Patient 3646/56 before and after operation (Figs 1 and 2).





(*2*) *Orbital Tumour.*—Patient 1485/57 before and one week after operation (Figs 3 and 4). This was found to be a cystic growth entirely in the muscle cone. *X*-rays showed an enormous expansion of the orbital margins and of the optic foramen without erosion. There was no pulsation on coughing, etc.

The histological report was inconclusive, but the pathologist thought the tumour must have been a haemangioma.

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OCULAR CASES OF MEDICAL INTEREST*

BY

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THE following cases were encountered some years ago while I was abroad on active service. This has unfortunately made it impossible to give absolutely complete reports or to make full acknowledgements to the service practitioners who sent in these cases for consultation.

Case Reports

Case 1. Myopathica Atrophica.—A yeoman of signals aged 22 years was admitted to the Military Hospital, Gibraltar, on August 31, 1943. He complained of having difficulty in facing the light for 10 days. The eyes ached but did not water, and had become bloodshot in the last 2 months, but especially in the last week when he attended the American hospital at Londonderry for conjunctivitis. Moreover, he had been losing weight for the last 6 months with much thinning of the arms, legs, and chest, more especially on the right side; the left arm gave more trouble when signalling (semaphore) and tired more easily; he lost control of the right leg occasionally. His face was much thinner and he was becoming "pot-bellied". He was sleeping well.

Past History.—The patient had been knocked unconscious at the age of 13 years and had sustained severe bruising of the forehead but no fracture of the skull. There was no amnesia for the period of the accident. He had spent 2 weeks in Shotley Naval Hospital for "bronchitis" in 1936. A right herniotomy had been carried out in 1938. He had had anti-syphilitic treatment in 1940 (fifty injections of N.A.B.) at Singapore. He had developed severe tonsillitis 8 months ago.

Family History.—His father was dead (heart trouble); his mother, who was alive, had been suffering from "some obscure muscular disease" for the last 20 years. There were two sisters, both alive and well.

Examination.—The patient had a drawn, haggard look with staring eyes, the bulbar conjunctivae being rather injected with darkish vessels. Facial muscular wasting was present with atrophic ectropion and weakness from wasting of the orbicularis oculi. The sternomastoids were wasted as well as the shoulder muscles. Organic reflexes normal.

The conjunctivae over both globes appeared thicker than normal, with thickened, dilated, tortuous, purple veins, and had a dry, greasy appearance. The arteries were also dilated but not so much as the veins. Photophobia but no lacrimation was apparently present. At no time did the patient give any sign of epiphora despite the atrophic ectropion and the engorgement of the conjunctival vessels. The pupil reactions were normal. There was no evidence of any intrinsic ocular muscle weakness. The bulbar appearance was that of a chronic episclero-conjunctivitis of a diffuse nature which was apparently secondary to exposure owing to the incapacity of the patient to close his lids properly over the globes. Other evidence of "exposure keratitis" was not seen.

The visual acuity was 6/6 without correction in both eyes. Both fundi were normal.

The case, being truly medical, was transferred to the physician, Major Leonard West, R.A.M.C., who reported on the general condition as follows:

Looks drawn and jaded. Wasting of facial musculature, both scapula and shoulder muscles affected. Right brachio-radialis affected, while there is also wasting of the right interosseus muscle. Right leg adductors and tibialis anticus wasted.

General appearance "pot-bellied" with some "winging" of the scapula on the right side.

* Received for publication August 12, 1957.

Drags the right foot but the power is good in both feet with the exception of the right on dorsiflexion. No evidence of tremor or fibrillation. Superficial and organic reflexes normal. No Rombergism. Deep reflexes difficult to elicit and responses sluggish. No gross scoliosis present. Throat, larynx, lungs, heart, and abdomen normal. Urine, no albumen or sugar. Kahn test negative. Patient writes with his right hand but is otherwise left-handed. Electrical reactions as follows: muscles of shoulder girdle normal; quadriceps and hamstrings normal; calf muscles and peronei normal; anterior tibial group no faradic reaction. Galvanic reaction fairly quick except in tibialis anticus, right and left, where it is almost absent.

Diagnosis.—Myopathic atrophy with exposure episclero-conjunctivitis.

Case 2. Processional Caterpillar (*Ctenocampa processina*) Hairs in the Eye.—An aircraftman aged 22 years got some cigarette ash in his left eye while watching and handling processional caterpillars on North Front, Gibraltar, some 24 hours previously in a very blustery wind. He admitted squeezing some of the caterpillars, but he had not touched his eye until some ash went into it, when he rubbed it with his fingers. He now felt that there was something in the eye still and came to have the "foreign body" removed.

Examination.—The conjunctiva over the left globe was very red and injected with considerable temporal chemosis; the area of chemosis was dimpled in the centre as if it had been burnt in that area. The dimpled zone was covered with a sticky viscid mucopus of a pearly-white appearance. The whole appearance resembled that of "pitting" when an oedematous surface is pressed with the finger. Circumciliary injection was present, suggesting iritis, but there were no keratic precipitates. The corneal limbus nearest to the swelling looked oedematous also. The fundus and lens were clear. The ocular tension was normal. There was no staining of the cornea.

The slit-lamp examination showed the true nature of the injury, as the pitted zone of the conjunctiva was full of golden barbs which were migrating into the corneal region near the limbus. There was a most severe conjunctival reaction about the barbs, and 10 days later they had migrated into the cornea where they became encapsulated in a white shroud—a protein-precipitation reaction—and looked like an Egyptian mummy in a sarcophagus. At a later stage, fibrous tissue enveloped the barbs, and the affected barb would become quiescent. As new barbs entered the cornea, so the reaction of the globe was maintained with acute exacerbations of the lesion. It seemed highly probable that these very minute barbs would ultimately penetrate the globe and destroy it. While he was under my care, keratic precipitates did not develop. The ultimate fate of this aircraftsman's eye is unknown as he was invalided home, but the prognosis is poor.

Case 3. Accidental Solanaceous Mydriasis.—A soldier in the Pioneer Corps, aged 22 years, complained that while he was squeezing a small, green "apple" some of the juice had entered both eyes, causing him great pain and making him "blind".

Examination.—Both globes showed marked congestion and injection of the bulbar conjunctiva. The lids were in a state of blepharospasm, and on raising the lids a considerable amount of lacrimatory fluid escaped from the conjunctiva, where it had been trapped by the spasm. The visual acuity was 6/24 in both eyes, and the patient was quite unable to read any print of reasonable size. The pupils were grossly dilated and inactive to light. The patient denied that he had had atropine put in them at the First Aid Post, and this was confirmed. The addition of correction for distance and of +3·00D sph. resolved the visual condition in both eyes.

Diagnosis.—Traumatic mydriasis was due to the accidental instillation of an alkaloid from an indigenous solanaceous plant recognized as Egyptian Henbane (*Hyocyamanus muticus*). This plant, which is well known in the Mediterranean zone, both East and West, bears a small, green fruit like an unripe tomato about 1·25 in. in diameter. It is called "Sakan" or "The Drunken", and among various other names given to the

fruit by the armed forces are "Gibraltar Tomato", "Monkey's Apple", and "Spanish Apple". It yields 83 per cent. of its alkaloids as *L. hyocyamine*, an 0·5 per cent. solution of which is equivalent to a 2 per cent. solution of atropine.

Case 4. Melanosarcoma of the Macular Area.—A Chief Petty Officer, R.N., aged 38 years, with 22 years' service, first attended the Military Hospital at Gibraltar on September 30, 1941, complaining of some impairment of reading during the last 4 weeks, with alteration in the shape of the letters, which appeared bent. He had always been a dead shot and had never had a squint or any ocular injury. He had been in perfect health, and had been through a number of important actions, and at sea most of the time. The right eye was the one that was giving trouble.

Examination.—He was a strong, healthy type, not nervous, introspective, or unduly sensitive, of good physique, quick and intelligent. The visual acuity in the right eye was 6/36 with +1·50 D sph., and 6/9 with +0·50 D cyl., 180°. The metamorphopsia could be corrected. In the left eye the visual acuity was 6/6 +0·75 D sph. and 6/5 with +0·50 D cyl., 85°. The pupil reactions were normal, the right cornea clear, and there were no other obvious abnormalities in the ocular media to affect the form of the retinal image. The right fundus showed "scattering" of the macular reflex, with heightened reflexes over the whole area. The macula lutea did not look abnormally coloured, but may have shown a slight increase in its normal yellowish appearance. The reflexes from the vessels as they emerged from the optic disc were much increased. The ocular tension of both eyes was normal.

X-rays of skull and orbits were negative, Kahn test negative, and physician's pathological report negative.

Progress.—The patient was told to report again in one month, but he went to sea on active service, and did not report again until January 19, 1942. He complained that objects appeared more bent than ever, and that he had difficulty in seeing clearly with the right eye for both distance and near. He had no other complaint.

The visual acuity in the right eye was 6/60 with +1·50 D sph., and 6/18 with +0·50 D cyl., 180°. It was not possible to correct the metamorphopsia.

The right fundus showed heightened reflexes, especially along the retinal arteries as they issued from the disc, where they almost resembled "pipe-stem" sheathing. The macular reflex was now absent, with some very slight disturbance of the pigment layer. The foveal pit had disappeared, and there seemed to be a potential detachment of the central area of the retina, or even a very small shallow detachment of this area which could not be seen ophthalmoscopically as there was no increase in the hypermetropia.

The ocular tension of the right eye was normal, anterior transillumination negative; visual fields showed relative scotomata for red, green, and blue, with ¼° and ½° targets.

The Kahn test was negative, the sinuses, teeth, and tonsils clear of infection, blood pressure 142/82, and urine clear, without sugar or albumen.

X-rays of the skull, right optic foramen, and right orbit were negative.

Further interrogation elicited the fact that the patient's mother had died from "malignant pigmented ulcers of the leg" some years previously.

Diagnosis.—A provisional diagnosis of very early melanosarcoma of the macular region was made. The patient was discharged to the U.K. for further opinion and enucleation of globe, and entered the Royal Naval Hospital, Gosport, where a report on the pathology of the globe later confirmed the diagnosis of melanosarcoma.

My sincere thanks are due to the Officer Commanding, Military Hospital, Gibraltar (1941–44), Col. W. Jackman, T.D., late Surg. Capt. P. N. Button, R.N., Medical Liaison Officer, Gibraltar Command, and Major Leonard West, R.A.M.C., and to the Royal Naval Oculist i/c, Gosport, 1941, for his kindness, despite the exigencies of war, in sending me a copy of the pathological report on Case 4.

Brit. J. Ophthal. (1958) **42**, 441.

APPLIANCE

REFRACTION IN MARKED PTOSIS AND BLEPHAROCHALASIS*

BY

J. D. MAGOR CARDELL

London

IN cases with marked ptosis or blepharochalasis it is not easy to obtain a sufficient area of pupil for refraction. The patient can sometimes assist by raising the lid or skin with a finger, but it may be difficult for him to avoid interference with the trial frame, and this difficulty is enhanced when the eyes are deep-set.

FIGURE.—Ptosis wire mounted on ring.

The appliance here illustrated (Figure)† is made to fit into the back cell of the frame and can be altered, by finger adjustment of the wire, to reach and lift the lid or skin to the desired degree. The wire must be so arranged that it does not press on the globe and produce an artificial astigmatism. The ring on which the ptosis wire is mounted is prevented from rotating by the pins set horizontally in its edge. It can be used for either eye and is oxidized to prevent reflections.

* Received for publication December 12, 1957.

† This appliance was made for me by Messrs C. Davis Keeler, 39 Wigmore St., London, W.1.

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BOOK REVIEWS

A Century of International Ophthalmology. By SIR STEWART DUKE-ELDER, G.C.V.O., 1958. Pp. 91; illustrated with portraits of past Presidents. Kimpton, London. (12*s.* 6*d.*).

The first International Congress of Ophthalmology was held in Brussels in 1857; 150 delegates attended from 24 countries and states (all but two from Europe) and 34 official papers were read. The XVII Congress was held in Montreal and New York in 1954; 2,164 delegates attended from seventy countries from all over the world, and more than 194 communications were published. These figures alone emphasize the vitality of the international life of ophthalmology, the story of which, together with those of the International Council and the Federation of Ophthalmological Societies, is fascinatingly told in this short book. The history of international co-operation mirrors not only the great fundamental advances in the specialty, but also the political troubles of the past one hundred years, explaining why the meeting to be held in September, 1958, is not the twenty-sixth of what was originally planned to be a four-yearly series, but only the eighteenth. This year, the centenary (as near as may be) congress is to be held very appropriately in Brussels, the city which first gave it hospitality. By no means its least attractive feature will be this book which explains so lucidly the background against which so many of us will be assembling, pictures what has been achieved in the past, and analyses the possible steps through which the next century may prove even more fruitful.

Principles of Ophthalmoscopy. By J. K. ERBAUGH. 1958. Pp. 69, 28 figs (8 in col.). Blackwell Scientific Publications, London. (42*s.*)

This little book is designed to aid the student, general physician, and neurologist to interpret ophthalmoscopic appearances. Difficulties which may arise because of opacities of the ocular media are discussed, and the normal and abnormal features of the optic disc and the retinal vessels are described. The book ends with a short description of the retinopathies secondary to vascular disease and such conditions as diabetes.

My Path in Science. By VLADIMIR FILATOV, trans. from the Russian by G. H. Hanna. 1958. Pp. 183, 68 figs. Foreign Languages Publishing House, Moscow (1957); Central Books, London. (3*s.* 6*d.*)

This is a small volume translated into English by G. H. Hanna from a book written by V. P. Filatov in 1955, the year before he died, and containing what the author considered to be the highlights of his contributions to ophthalmology. These are dealt with in eight chapters, the subjects of which include the tube-flap graft, glaucoma, trachoma, the use of saliva instead of tears, the treatment of seborrhoea of the Meibomian glands, first-aid in eye injuries, corneal grafting, and tissue therapy. The last two are the most important. Filatov's contributions to the technique of corneal grafting were undoubtedly great, for it was he who introduced the use of cadaver eyes and thereby made possible the popularization of the method. The value of his technique of tissue-therapy is more problematical. It will be remembered that, having observed the clearing of an opaque cornea in the neighbourhood of a graft, he claimed that all living tissue, human, animal, or vegetable,

when isolated from the organism and retained under conditions that are unfavourable but not lethal, produces "biogenic stimulators", non-specific substances which have the property of stimulating vital processes in the organ into which they have been introduced. It is quite possible that there is some biological truth in this concept and that a tissue in difficulties produces stimulants which help to preserve its viability and which in turn may similarly stimulate a diseased organ into which they are introduced. It is interesting that Filatov himself agreed that the substances themselves are unknown and that their action appears to be quite non-specific. At the same time, there would be few who would agree that they have any dramatic value in the treatment of diseases such as those in which Filatov himself claimed to have produced a marked therapeutic effect—myopic degeneration, optic atrophy, primary pigmentary degeneration of the retina, and so on. The present volume, however, gives in a simple and concise form his final views on a subject which has excited considerable interest.

Ocular Anatomy. By J. D. SPOONER. 1957. Pp. 226, 216 figs. Hatton Press, London. (45*s.*).

This book presents a clear and concise account of the structure and development of the eye and its related structures.

It is written primarily for students of ophthalmic optics, and is designed, therefore, for those who do not possess any background knowledge of general anatomy.

There are several minor errors in the text, and some of the clinical conclusions which are derived from anatomical facts are incorrect, but on the whole the book represents an accurate, non-controversial account of present-day concepts of basic ocular anatomy.

Year Book of Ophthalmology (1957–1958 Series). Edited by DERRICK VAIL. 1958. Pp. 423, 90 figs. Year Book Publishers, Chicago. ($7.50; 47*s.*)

There is now some hope for the world, for the Americans are learning sense; they have at long last realized the relative importance of ophthalmology and otorhinolaryngology. This year, for the first time, there is a *Year Book of Ophthalmology* instead of the *Year Book of Eye, Ear, Nose and Throat* with which we have had to put up for more than half a century. The result is a more thorough appreciation of ophthalmological progress during the last year, a more useful and interesting book, and a book ophthalmologically pure, without contamination.

The volume is introduced by a long special article on recent advances in ocular therapeutics written by Irving H. Leopold. This is an excellent article which contains a vast amount of information on the treatment of many ocular conditions and includes methods of treatment as different as the chemotherapy of cancer on the one hand, and the use of tranquillizers on the other. The remainder of the book, compiled by Derrick Vail, abstracts the more important papers on all aspects of ophthalmology which have appeared during the year under review, and contains more than twice the number of articles and editorial comments that have been included in the past. The articles are well chosen, their abstraction is full, and the comments—as usual—are short, fearlessly critical, and unusually useful. To the ophthalmologist who wishes to revise or supplement his reading in the current journals, the book will be found indispensable.

Management of Complications in Eye Surgery. Edited by R. M. FASANELLA. 1957. Pp. 422, 23 figs, 23 tables, bibl. Saunders, London. (112*s.*)

In the chapter on complications of cataract surgery Robb MacDonald writes: "The only surgeon who does not have to deal with complications is one who does not operate". Nevertheless, a book of 422 pages devoted to the management of complications in eye surgery may strike the ophthalmologist as somewhat horrifying until it is realized that

management is taken to include prevention and much of the discussion concerns prophylaxis. Because the surgical handiwork of colleagues always tends to look better than one's own, it is indeed comforting to find that surgical snags are universal. The book is full of practical advice given by American surgeons of wide experience.

The first three chapters discuss anaesthetic, medical, and psychiatric complications and they are particularly interesting to the ophthalmic surgeon because the responsibility of these aspects of treatment are usually shared with colleagues. There follow chapters on purely ophthalmic subjects: intra-ocular foreign bodies, ptosis, lacrimal surgery, keratoplasty, iris surgery and trauma, glaucoma operations, cataract, extra-ocular muscle surgery, retinal detachment, orbit and lid surgery, and endocrine exophthalmos. The last two chapters discuss rehabilitation of the patient with low visual acuity and the estimation of loss of visual efficiency.

A book which is so practical and explains so many small but important points in technique is not easy to review. It has been written to serve as an advanced reference book and its object has certainly been achieved. It should find its way into the hospital library wherever ocular surgery is practised, and it will pay dividends to the trainee who cares to thumb its pages before he undertakes an operative procedure which is new to him. It is well illustrated and the references for wider reading are chosen with discretion.

Veterinary Ophthalmology. R. H. SMYTHE. 2nd ed., 1958. Pp. 379, 59 figs. Baillière, Tindall and Cox, London. (42*s*.)

The appearance of the second edition of this book within 2 years of its first publication suggests that it is fulfilling a deservedly useful role. As would be expected after so short an interval, there are no radical changes. The 21 extra pages are occupied by a revision and rearrangement of quite a number of sections and the addition of new figures—all of which are improvements on those included in the first edition.

NOTES

OPHTHALMOLOGICAL SOCIETY OF THE UNITED KINGDOM

Annual Congress, 1958

The 78th Annual Congress of the Ophthalmological Society of the United Kingdom, held at the University College of South Wales and Monmouthshire, Cathays Park, Cardiff, on April 17 to 19, 1958, under the presidency of Mr. J. J. Healy, was attended by over one hundred members of the Society and several distinguished foreign guests.

The meeting opened with a discussion on the complications and treatment of the hypermature cataract. Mr. G. T. W. Cashell (*Reading*) approached the subject historically, describing the various complications of hypermature cataract that had gradually achieved recognition, secondary glaucoma and uveitis being the most important. He described how an anterior hypermature lens may prevent glaucoma and the special difficulties involved in extracting such a lens, and then considered the uveal reactions from escaping Morgagnian fluid, whether phaco-anaphylactic toxic or anaphylactic. Lastly, he dealt with rupture of the zonule with anterior or posterior dislocation of the hypermature cataract.

Mr. C. A. G. Cook (*London*) then considered and illustrated the pathological aspects of these complications, which he described under two main headings: the first comprising complications leading primarily to the production of glaucoma, and the second including conditions in which the primary pathological change is of an inflammatory nature which may or may not be responsible eventually for a secondary increase in intra-ocular pressure.

Phacolytic glaucoma, hypermature intumescent lens, and hypermature dislocated lens were representative of the first group, whilst the second comprised the so-called lens-induced uveitis, which was subdivided into phacotoxic uveitis and endopthalmitis phaco-anaphylactica.

Mr. J. R. Hudson (*London*) discussed the clinical recognition of the mature and hypermature cataract and its indications for surgical treatment. The hypermature cataract did not, he considered, present the problem that it previously presented, as cases were less frequently seen than in the past, and its complications were rarely encountered. He then described the varieties of hypermature cataract with their operative treatment, and detailed the clinical features of the three chief complications: phacolytic glaucoma, phacotoxic uveitis with secondary glaucoma, and endophthalmitis phaco-anaphylactica. He felt that the recognition of the early signs of hypermaturity should, in the absence of any general contraindication, be taken as a positive reason for advising operative treatment of a cataract, in order to prevent the development of these complications. The paper concluded with three illustrative cases.

Mr. A. Stanworth (*Manchester*) described how conjunctival fibrosis after a filtration operation could lead to drainage of the aqueous into a circumscribed filtration bleb, and how poor absorption from this bleb could then result in a recurrence of intra-ocular pressure. He described cases that illustrated this condition and discussed its cause and rectification.

Mr. S. J. H. Miller (*London*) discussed the origin and treatment of Fanconi's syndrome with band-type calcium deposits in the cornea. He described in detail a recent case of Fanconi's syndrome which had presented pseudo-fractures and was treated with calciferol. The calcium deposits that had developed within the cornea were then dispersed by maintaining the patient in a state of mild acidosis.

Dr. Maurice E. Langham (*London*) reported his experiments showing the effect of Neptazane on the intra-ocular pressure in the rabbit and in man. Neptazane was a new inhibitor of carbonic anhydrase and diffused into the eye more readily, while leaving the body more slowly, than Diamox. He found that, when Neptazane was given intravenously, both the intra-ocular pressure and the rate of formation of the aqueous decreased rapidly.

Mr. P. L. Blaxter (*Manchester*) described some cases of rhabdomyosarcoma of the orbit, including that of a boy who was still living 3 years after the removal of the growth. He discussed the clinical features and management of such cases in the light of previous reports.

Mr. R. L. S. Smith (*Manchester*) described the pathological aspects of Mr. Blaxter's cases.

A discussion on the role of the toxoplasma in ocular disease was opened by Prof. C. P. Beattie (*Sheffield*). He briefly described the biological characters of the toxoplasma and methods used to demonstrate toxoplasma infection. This protozoon had a singularly wide zoological and geographical distribution, with human infection varying from 5 per cent. in Iceland to 70 per cent. in Tahiti. It increased with age, and in the United Kingdom about one-quarter to one-half of the adult population had been infected, although the route through which it normally spread was still unknown. In the congenital form, where at least the placental rate of transmission is definite, a chorio-retinitis was the most constant manifestation. Most toxoplasmic chorio-retinitis was of congenital origin while the acquired infections were usually innocuous, featuring only a lymphadenopathy resembling glandular fever. The assessment of toxoplasmic antibodies was difficult in congenital cases since they had normally fallen to levels around those usually found in the general population as a result of latent infection; and one could at best only assess the probability of a toxoplasmic origin of any particular chorio-retinitis on the basis of serological tests. In young children, the diagnosis was relatively easy for, in the rare cases where antibodies are present, they were also generally of high titre.

Dr. J. K. A. Beverley (*Sheffield*) continued the discussion on toxoplasmosis by outlining the behaviour and growth of the organism in both non-immune and immune animals, giving evidence that even in highly immune animals multiplication could and did occur. He summarized the chemotherapy of acute experimental infections due to toxoplasma cysts, and discussed the hypersensitivity reactions and how they could best be controlled.

Mr. E. S. Perkins, Reader in Ophthalmology at the Institute of Ophthalmology, London, considered the more specifically ophthalmological aspects of toxoplasma infection, underlining that there was now good statistical, therapeutic, and pathological evidence that toxoplasmosis might cause uveitis in adults as well as a congenital chorioretinitis. Unfortunately, serological evidence of infection with toxoplasmosis was common in normal adults, and though a negative dye test probably excluded toxoplasmosis, a positive test did not prove the ætiology of an individual case of uveitis. It was possible that some cases of anterior uveitis were due to toxoplasmosis but those cases could not yet be recognized clinically. Cases of posterior uveitis were found to have a high percentage of positive dye-tests—particularly the acute and sub-acute groups. In a series of 106 cases of posterior uveitis out of a total of 653 cases of all types of uveitis, the acute and sub-acute posterior groups had 82 per cent. positive dye-tests (over 1:4) compared with some 50 per cent. in all other types of uveitis. As the patients in these groups were younger than the series as a whole, this made the higher incidence of positive tests even more surprising, for in the population as a whole the percentage of positive reactions was lowest in the younger age groups. Such cases were characterized clinically by focal patches of chorio-retinitis situated near the main retinal vessels or the macula, or at the margin of the optic disc. In many cases the active lesion occurred at the edge of an old scar, suggesting that most lesions were recurrences of congenital infections. There were, however, nineteen cases with a positive dye-test (including five with the lesion at the macula) in which there was no evidence of a previous lesion. Although it was difficult to prove that all or any of these cases were due to toxoplasmosis, it seemed possible that some followed infection acquired after birth or perhaps in adult life. Disseminated choroiditis was not associated with positive dye-tests and was unlikely to be due to toxoplasmosis; the results in retinal vasculitis also suggested that toxoplasmosis was not a common cause of that condition.

A discussion on pre-cancerous melanosis of the conjunctiva was opened by Mr. J. J. Healy (*Llanelly*), President of the Society. He described three recent cases illustrating different aspects of the condition, one of them with a frankly cancerous development. Dr. M. Lederman (*London*) discussed the role of radiotherapy in cancerous and pre-cancerous melanosis. His results of treatment at the Royal Marsden Hospital were assessed and the indications of such treatment were found to be directly at variance with those recently enunciated by Mr. A. B. Reese. Twelve out of fourteen patients with cancerous melanosis received radiotherapeutic treatment; of these two died of metastases and the remainder had survived for intervals of up to 7 years, so that it could be concluded that some of these tumours, at any rate, were in fact, radiosensitive. He had tried various methods of irradiation, including *beta*-irradiation from radium or radioactive strontium, and *gamma*-irradiation from radon seeds and radioactive tantalum wire; some cases were illustrated with an unduly severe reaction following the use of radioactive tantalum wire, and he concluded that *beta*-irradiation was to be recommended because of its minimal complications.

Mr. Frederick Ridley (*London*) described in detail the special contact shells used in this form of therapy, and Dr. A. L. Wells (*Llanelly*) described the pathology of certain cases of pre-cancerous melanosis, and compared them with similar lesions in cases of cutaneous squamous epithelium.

Dr. R. W. Harrington (*Glasgow*) discussed the damage to the chiasma and hypothalamus which might follow the use of radioactive implants in the pituitary fossa in treating

inoperable hormone-dependent neoplasia. He described how these seeds were accurately inserted, using the isotope yttrium-90 as a short-acting *beta*-emitter in preference to the *gamma*-emissions of radon. The field changes found in twelve patients (out of a total of 63 examinations) were illustrated and some of the pathological material was described.

Mr. Arthur Lister (*London*) presented observations on the results of perforating cyclodiathermy, which he found more effective than surface cyclodiathermy, especially in the treatment of chronic glaucoma in the elderly patient, although he had some doubt as to the permanence of its effect.

Mr. A. G. Cross (*London*) discussed the treatment of chronic uveitis, which has been revolutionized by the use of the adreno-cortical hormones, which readily damped down the acute cases until they had run their usual short and pre-determined course. He discussed in detail the various methods of dealing with the more serious problems of chronic uveitis, the outlook of which, although much better than it had been 10 years ago, was still often grave, and frequently resulted in blindness.

Mrs. Dorothy Miller (*London*) reported a case of anorexia nervosa in a young woman, which was followed by the development of subcapsular cataracts thought to be nutritional in origin.

Dr. John McArthur (*London*) demonstrated a microscope that he had developed for use in clinical and research work, which was only the size of a miniature camera.

Mr. Dermot Pierse (*Croydon*) described his method of iris inclusion. He felt that, in many cases, particularly of early shallow-angle glaucoma, the classical complete iridectomy with incarceration of one or both iris pillars was unnecessarily severe and mutilating. He had accordingly developed a method of incarcerating the peripheral iris without destroying the sphincter, by introducing forceps laterally from a slightly oblique *ab externo* incision.

Mr. M. J. Gilkes (*Brighton*) reported on his recent investigation of trachoma in Jordan and Gambia. He had found a definite variation from the incidence figures previously reported, and also differences in the clinical characteristics and sequelae of the disease, which appeared to be associated with variations in the incidence of the H.P. inclusion bodies.

Mr. Redmond Smith (*London*) described his analysis of the incidence of various types of glaucoma at the St. Mary's Hospital Glaucoma Clinic, Western Ophthalmic Hospital, London. After detailing his methods, he concluded that closed-angle glaucoma was, in fact, much more common than simple glaucoma.

Mr. J. A. E. Primrose (*London*) described the value of the Priscol test in glaucoma. This test produced a small but transient increase in normal eyes, but in fifty cases of chronic simple glaucoma there was a mean rise of tension of 12·2 mm. Hg, while most cases of narrow-angle glaucoma gave a normal response. The Priscol effect was probably due to its effect on the vessels because of an increased formation of aqueous humour.

Surgeon Commander D. P. Gurd, R.N., emphasized the importance of lime burns in industry and described three recent cases affecting the external eye. He discussed the results of some experimental studies on rabbits, showing the influence of sub-conjunctival Priscol and topical hydrocortisone ointment.

Dr. H. Pines (*London*) gave a pictorial demonstration based on the work of Krückmann, which suggested that Rouget cells were absent from the retinal capillaries.

Mr. T. Keith Lyle has been appointed examiner for the final F.R.C.S. in Ophthalmology of the Royal College of Surgeons of England.

Mr. James H. Doggart and Mr. Patrick Trevor-Roper have been made honorary members of the Peruvian Ophthalmological Society. Mr. Doggart has also been made an honorary member of the Oto-neuro-ophthalmological Society of Argentina.

Brit. J. Ophthal. (1958) **42**, 448.

OBITUARY

William Halliburton McMullen

William Halliburton McMullen was born on July 19, 1879, the son of a South London general practitioner, Dr. William McMullen. He was educated at the City of London School and King's College, London, where as a medical student he won first and second year scholarships, and was assistant demonstrator of ophthalmology and prosector. He continued his clinical work at King's College Hospital, qualifying in 1898 and taking his M.B., B.S., with Honours in 1900. He held the posts of junior and senior house surgeon and took the F.R.C.S. (England) in 1900. He had now become interested in ophthalmology and became house surgeon at the Royal Westminster Ophthalmic Hospital, and was later appointed pathologist and curator. At almost the same time he became assistant ophthalmic surgeon to the Miller General Hospital and shortly afterwards, in 1906, assistant surgeon to the Royal Westminster Ophthalmic Hospital. From this time his career as an ophthalmic surgeon really began. He was for 8 years ophthalmic surgeon to the Hospital for Sick Children, Great Ormond Street, and for 4 years to the Royal Northern Hospital. During the 1914–1918 war he was ophthalmic specialist to the Central London Recruitment Depot, and in 1918 was awarded the O.B.E. In 1929 he was appointed honorary ophthalmic surgeon to Charing Cross Hospital. He maintained his connexion with the Royal Westminster Ophthalmic Hospital, wherein he was senior surgeon from 1927–1936, and when he retired from the staff he still maintained contact through the House Committee. He was President of the Section of Ophthalmology of the Royal Society of Medicine in 1937–1938, and also a Vice-President of the Ophthalmological Society of the United Kingdom.

In 1936 he retired from his active hospital work at the age of 60, but in 1939 he took up an appointment with the Emergency Medical Service. After 1945 he continued to take a great interest in ophthalmology and was an original member of the Faculty of Ophthalmologists. During his career he contributed many papers to the literature.

He was a man of most distinguished appearance, which the early whitening of his hair tended to enhance. Very quiet and reserved in manner, he yet had an authority which all respected. He was a very skilful, painstaking surgeon and an acute diagnostician, and his learning and experience were always at the service of his assistants and colleagues. He was beloved by his patients, with whom he was ever kind and gentle, reassuring them by his calm, quiet manner. He was a very competent artist in landscape work, both with pencil and in water-colour, and derived much pleasure from this, though he was very modest about it and could not be persuaded to exhibit. Those who saw his work, however, had the highest praise for it. He was fond of a game of golf, and up to 1939 was a member of Oxhey Golf Club, where he would sometimes entertain his friends.

In 1906 he married Miss Kate Constance Higgins, who survives him with three sons, and to them the sympathy of all his many friends, ophthalmologists and others, will go out.